An epidemiological evaluation of the UK screening programme for congenital dislocation of the hip

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Ph. D. thesis

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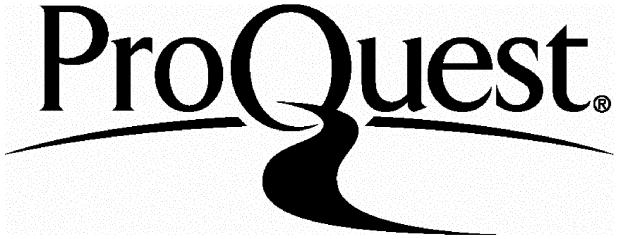

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ABSTRACT

An epidemiological evaluation of the UK screening programme for congenital dislocation of the hip

In the United Kingdom (UK), a policy of universal clinical screening was formally adopted in 1969. This screening programme was established to detect infants with hip instability so that early conservative treatment could be applied, and congenital dislocation of the hip (CDH) averted. However, the effectiveness of clinical screening has not been evaluated in a randomised trial, and is controversial. National data on effectiveness are lacking and individual centres have reported conflicting results. Although ultrasound imaging of the newborn hip has been adopted as a universal primary screening test in some European countries, enthusiasm for its introduction in the UK has been tempered by the consequent fourfold increase in the number of children identified as requiring early treatment.

In this thesis the scientific basis of the current UK screening policy for CDH and alternative strategies is critically appraised, and a national study undertaken to evaluate the existing programme presented. Current screening and management practices were characterised from data obtained from a national postal survey of all maternity units in the UK and Irish Republic. Paediatricians were asked to report children in whom they initiated abduction splinting through the British Paediatric Surveillance Scheme, which operates through an active surveillance scheme. Children in whom abduction splinting was initiated or who received a first operative procedure for CDH were reported through a similar scheme established among orthopaedic surgeons. Reported cases of a first operative procedure were validated with Hospital Episode System data for 18% of births.

The extent of under ascertainment of cases was estimated by capture-recapture analysis.

The prevalence of a first operative procedure for CDH was 0.78 per 1000 live births (95% confidence interval (CI): 0.72, 0.84) and of conservative treatment, 1.6 per 1000 live births (95% CI: 1.4, 1.7). More than two-thirds of the children who received a first operative procedure had not been detected by screening, while conservative treatment had been unsuccessful in a fifth. In the last decade there has been a fivefold increase in the use of ultrasound imaging, which in 1994, was used in 69% of maternity units as a secondary screening test for infants at high risk of CDH or with clinical hip instability. In this first national study, the prevalence of surgery was found to be similar to the prevalence of CDH before screening was introduced, although twice as many infants received conservative treatment than previously were affected.

CONTENTS

1: Introduction 13

2: Literature review 14

2.1 Screening: definition and terminology 14

2.2 Background 19

2.2.1 Definition and terms 19

2.2.2 Aetiology 20

2.2.3 History 21

2.2.4 Clinical screening examination 22

2.2.5 Current UK Policy 23

2.3 Screening and CDH 25

2.3.1 Is CDH an important public health problem? 25

Sequelae of untreated CDH 25

Treatment of established CDH 26

Prevalence 29

Prevalence of CDH before screening was introduced 29

2.3.2 Is there a recognisable latent or early symptomatic phase of CDH? 31

Prevalence of NHI 31

2.3.3 Is there a suitable test? 32

Risk factors for NHI 33

Continued diagnosis of clinically presenting CDH 34

2.3.4 Is early conservative treatment effective? 34

2.3.5 How effective is the screening programme for CDH? 37

Measuring outcome 37

Natural history 45

Failures of early treatment 45

Economics 47

2.4 Ultrasound examination of the newborn hip 48

2.5 Conclusion 50

3: Methods 52

3.1 National survey of screening and management 52

3.1.1 Survey respondents 52

3.1.2 Questionnaire design 53

3.1.3 Pretesting the questionnaire 56

3.2 Surveillance study 57

3.2.1 Introduction 57

3.2.2 Routine data sources 57

3.2.3 History of surveillance 59

3.2.4 Definition of surveillance 59

3.2.5 Requirements of a surveillance scheme 60

3.2.6 Ethical considerations 61

3.2.7 The British Paediatric Association Surveillance Unit 62

3.2.8 Case definition and study period 63

3.2.9 Development and maintenance of the orthopaedic reporting base 65

Development 65

Maintenance 65

3.2.10 Surveillance methods 66

Case notification 66

Encouraging response 67

3.2.11 Validation of the orthopaedic and paediatric reporting bases 68

3.2.12 Follow up of notified cases 69

3.2.13 Pretesting the follow up forms 70

3.2.14 Data entry and coding of forms 71

3.3 Validation of case ascertainment 73

3.3.1 Background 73

3.3.2 Capture recapture analysis 73

3.3.3 Validation of the cases of abduction splinting 74

3.3.4 Validation of the cases of a first operative procedure 74

3.3.4.1 Validation areas 74

3.3.4.2 Identification of cases of a first operative procedure from HES data 75

3.3.4.3 Case definition 76

3.3.4.4 Preparation of HES data 77

3.3.4.5 Ethical approval 77

3.3.4.6 Design of validation forms 78

3.3.4.7 Training 78

3.3.4.8 Data storage and entry 79

4: Results 81

4.1 Survey of screening and management 81

4.1.1 Response 81

4.1.2 Screening by clinical examination 82

4.1.3 Use of ultrasound 83

4.1.4 High risk infants 85

4.1.5 Management 85

4.2 Surveillance study 88

4.2.1 The orthopaedic reporting base 88

4.2.2 The paediatric reporting base 90

4.2.3 Compliance with the reporting schemes 91

4.2.4 Cases of abduction splinting 95

Prevalence 95

Characteristics of confirmed cases of abduction splinting 96

Geographical distribution of cases of abduction splinting 98

Mode of presentation of cases of abduction splinting 99

Diagnostic tests prior to abduction splinting 100

Treatment of NHI 100

4.2.5 Cases of a first operative procedure 101

Prevalence 101

Characteristics of confirmed cases of a first operative procedure 103

Geographical distribution of cases 104

Seasonality 106

Hip affected 108

Clinical examination 108

Imaging 108

Mode of presentation 110

Age at first operative procedure 110

Prior splinting 112

Diagnostic tests 112

Complexity of first operative procedure 112

4.2.6 Children treated for CDH in the Republic of Ireland 113

Child who received a first operative procedure 114

Children who received abduction splinting 114

4.3 Validation 115

4.3.1 Validation of cases of abduction splinting 115

4.3.2 Validation of cases of a first operative procedure: HES data 116

4.3.3 Source by which children identified, by region 117

4.3.4 Effectiveness of search strategies 118

4.3.5 Ascertainment-adjusted estimate of the prevalence of a first operative procedure 119

4.3.6 Comparison of the characteristics of cases by source of identification 120

4.3.7 Variable catchability 122

4.3.8 Application of results of the validation study to national figures 122

5: Discussion 124

5.1 Survey of screening practices 124

5.1.1 Potential sources of bias 124

5.1.2 Comparison to previously published data 126

5.1.3 Performance and reporting of ultrasound imaging 128

5.1.4 Management of infants with a presumptive positive screening test 129

5.1.5 Identification of infants at high risk of CDH 131

5.2 Surveillance study 132

5.2.1 Sources of bias 132

5.2.1.1 Completeness of the OS and BPASU reporting bases 132

5.2.1.2 Return of cards and forms 133

5.2.1.3 Under ascertainment of cases 136

5.2.2 Comparison to the published literature on prevalence of a first operative procedure 138

Before screening was introduced 138

After screening was introduced 139

5.2.3 Characteristics of infants and children who received a first operative procedure in the current study 142

5.2.4 Complexity of surgery 144

5.2.5 Prevalence of abduction splinting: comparison to the published literature 146

5.2.6 Characteristics of reported cases of abduction splinting 150

6: Conclusion 151

6.1 Summary of findings 151

6.2 Implications for the current screening programme 152

6.3 Implications for potential routine surveillance of the screening programme 158

6.4 Implications for clinical practice 159

6.5 Implications for future research 159

References 162

Acknowledgements 179

Tables

Table 2.1 Wilson and Jungner's criteria for an appropriate screening programme 18

Table 2.2: Summary of current UK recommendations [reproduced from 1986 SMAC report][1] 25

Table 2.3 Prevalence of a first operative procedure for CDH 39

Table 3.1 Coding frame for means of detection of children who received a first operative procedure 72

Table 3.2 Coding frame for means of detection of children who received abduction splinting 73

Table 3.3 Classification of children grouped according to means of case ascertainment 80

Table 4.1 Use of ultrasound 84

Table 4.2 Criteria used to identify infants at high risk of CDH. 85

Table 4.3 Source from which surgeons were identified for inclusion in the orthopaedic surveillance scheme 90

Table 4.4 Reported special interest(s) of paediatricians not included in the BPASU scheme during the study period 91

Table 4.5 Reasons for ineligibility for notified cases of abduction splinting 95

Table 4.6 Characteristics of confirmed cases of abduction splinting 97

Table 4.7 Reported risk factors for confirmed cases of abduction splinting 98

Table 4.8 Geographical variation of confirmed cases of abduction splinting 99

Table 4.9 Reasons for ineligibility of reported operative cases 102

Table 4.10 Characteristics of confirmed cases of a first operative procedure 103

Table 4.11 Prevalence of known risk factors of cases of a first operative procedure reported to the Orthopaedic Surveillance Scheme 104

Table 4.12 Geographical distribution of cases 106

Table 4.13 Complexity of the first operative procedure 113

Table 4.14 Background characteristics of children treated in the Republic of Ireland . 115

Table 4.15 Reasons for ineligibility of children identified by HES data 117

Table 4.16 Source by which children identified, by region 117

Table 4.17 Under ascertainment-adjusted prevalence of a first operative procedure, by source 119

Table 4.18 Geographical variation in the ascertainment-adjusted prevalence of a first operative procedure 120

Table 4.19 Comparison of characteristics of children identified by Hospital Episode System data (HES) only and those ascertained through the Orthopaedic

Surveillance (OS) Scheme 121

Table 4.20 Complexity of first operative procedure, by source 122

Table 5.1 Estimates of prevalence of CDH after screening was introduced: comparison to the literature by relevant region or county 141

Figures

2.1 Test result and disease status 15

2.2 Association of study size and estimates of the prevalence of late-detected CDH, UK 42

2.3 Prevalence of late-detected CDH and length of follow up of cohort 44

4.1 Percentage of maternity units in the UK and Irish Republic with access to ultrasound imaging, by year of introduction 84

4.2 Frequency of splint types used in UK maternity units, 1994 87

4.3 Monthly card return rate of Orthopaedic Surveillance scheme 92

4.4 Percentage of reporting cards returned, by country or former NHS region 94

4.5 Distribution of reported cases of a first operative procedure by month of birth . . . 107

4.6 Cases of a first operative procedure: side affected 109

4.7 Distribution of age at first operative procedure 111

5.1 Secular trends in the prevalence of abduction splinting in four UK centres, 1960-1989 145

5.2 Prevalence of abduction splinting in relation to policy of immediate treatment for neonatal hip instability, by region 147

5.3 Prevalence of abduction splinting and a first operative procedure for late-detected CDH, by region 149

Appendices

2.1 current and past membership of the Medical Research Council Working Party . . . 180

3.1 questionnaire, survey of screening and management practices 181

3.2 covering letter, survey of screening and management practices 188

3.3 protocol card sent to paediatricians participating in the BPASU 189

3.4 protocol card sent to orthopaedic surgeons participating in the OS scheme 190

3.5 article published in the *British Orthopædic News,* spring 1993 191

3.6 reporting card sent to orthopaedic surgeons 192

3.7 advance notification to paediatricians of CDH study 193

3.8 questionnaire sent to surgeons identified by manpower census 194

3.9 questionnaire sent to paediatricians identified by manpower census 195

3.10 follow up form for notified cases of abduction splinting 196

3.11 follow up form for notified cases of a first operative procedure 197

3.12 questionnaire to identify potential sources of abduction splinting 198

3.13 validation form 199

1: INTRODUCTION

Congenital dislocation of the hip is a potentially disabling condition which affects approximately 1 in 1000 children in the United Kingdom. Universal screening by clinical examination was introduced more than twenty-five years ago in the expectation that it would reduce, if not eliminate, through early detection and conservative treatment, the need for invasive surgery. The outcome of the current screening programme has proved controversial, since some individual centres have reported a reduction, some no change and others an increase in the number of children requiring surgery, although more children have received conservative treatment than would be expected to later develop CDH.

Universal primary screening using ultrasound imaging of the hips, which provides otherwise unavailable information on hip morphology, has been advocated. However, its introduction in other countries has been associated with an increase in the number of children receiving conservative treatment. The failings of the programme based on screening by clinical examination attest to the problems of introducing screening before recognised criteria for screening programme are met. The emergence of ultrasound imaging provided the impetus to evaluate the current programme and the potential alternatives.

In this thesis, an epidemiological evaluation of the current national screening programme for CDH is described, the first such evaluation to be carried out on a national basis. In chapter 1, an historical account of the current programme is presented, with a critical appraisal of the scientific basis underlying the current recommendations for screening. Each aspect of the current programme is reviewed in the light of the published literature.

In Chapters 2 and 3, respectively, the methods and results of a national study undertaken to establish current screening and management practices for CDH in the UK and Irish Republic, and to establish the national prevalence of treatment, both conservative and surgical, are described. The former was achieved by postal questionnaire sent to paediatricians responsible in every maternity unit in the UK and Irish Republic. The latter was undertaken by means of active reporting through an established national surveillance scheme among paediatricians, and a similar scheme established in collaboration with orthopaedic surgeons specifically for this study. The reporting bases of the two surveillance schemes were validated by comparison to manpower census data. Ascertainment of cases receiving a first operative procedure was validated with Hospital Episode System data for 18% of births. The findings of the study are reviewed in the light of published data and the implications for future research are discussed in Chapter 5.

2: LITERATURE REVIEW

2.1 SCREENING: DEFINITION AND TERMINOLOGY

Wald[2] has defined medical screening as "... the systematic application of a test or inquiry, to identify individuals at sufficient risk of a specific disorder to benefit from further investigation or direct preventive action, among persons who have not sought medical attention on account of symptoms of that disorder." Special ethical considerations apply to screening since it is preventive rather than curative medicine. Those screened are healthy individuals who are approached by the medical profession and there is an obligation not to initiate any action unless the full consequences of this are known. In contrast, symptomatic patients are usually seeking help and the

obligation is to treat them in the best way possible, even if there is incomplete knowledge about the disease and its remedy.[3] The screening of children, and especially of neonates, may be a more complex issue, since consent is sought from parents on a child's behalf.

Screening discriminates two groups of individuals based on a screening examination, those who are test positive and those who are test negative. Each group can be subdivided according to whether the test correctly allocates disease status (Figure 2.1).

Figure 2.1 Test result and disease status

		Disease status	
		+	-
Test result	+	true positives	false positives
	-	false negatives	true negatives

The ability of a test to designate people with disease as positive is called the *sensitivity* of the test or the *detection rate,* and the ability of a test to designate those who are not diseased as negative is called the *specificity*. Sensitivity and specificity can be varied reciprocally according to the level at which a test result is considered positive. If sensitivity and specificity change with age, the optimal time to screen is when the distribution of test results for affected and unaffected individuals has the smallest overlap, i.e. when the test is most sensitive and specific. Whereas poor sensitivity may reduce the effectiveness of screening as a preventive process, poor specificity may result in large numbers of false-positive diagnoses. The latter may lead to increased demand for diagnostic or therapeutic services, where available, and

potentially to over treatment, which may be high in human and financial costs.

The *predictive value* of a positive test is the probability that an individual is affected, given a positive test result, but this measure can also be expressed as the odds ratio of the number of affected to the number unaffected among those with positive results. The latter may provide a better indication of the reliability of a test. For example, a better technique might increase the positive predictive value from 95% to 98%. This appears to be a marginal improvement but the odds ratio is now 50:1 rather than 20:1. The proportion of a population that has detectable preclinical disease (or prevalence) is an important determinant of predictive value. Two tests for diseases of differing frequency may be equally sensitive and specific but the predictive value of the test for the less prevalent disease will be lower and hence the number of false positive test results higher. Thus, screening a large population in which few individuals are affected is less likely to be worthwhile. Prevalence itself depends on incidence, the rate at which new cases appear; on the rate at which cases disappear, through death or remission or cure; on the average length of the preclinical phase; and on whether the population has been screened previously.

The *reliability* or *reproducibility* of a test is its capacity to give the same result, positive or negative, whether correctly or incorrectly, in the same person at the same level of disease. This depends on the variability of the manifestation for which the screener is testing, and on the variability in the method of measurement and the skill with which it is made. Both intraobserver variation and interobserver variation should be low. Although a reliable test is not necessarily sensitive or specific, an unreliable test is unlikely to be sufficiently sensitive or specific to be useful. Estimating

variation between and within observers may be difficult if an individual cannot be examined on multiple occasions. For example, it has been suggested that repeated examinations may increase the propensity of an initially normal neonatal hip to dislocation, and should be avoided.[4,5]

The *lead time* is the interval from detection to the time at which diagnosis would have been made without screening. For a screening programme to be effective in reducing morbidity, there must be sufficient lead time in a sufficient number of screen-detected cases as well as an effective treatment which can be begun in this time. For example, a CDH screening programme may not be justified if it prevents established CDH in only a small proportion of affected children, or detects CDH too late for earlier treatment to be effective. Lead times cannot be calculated for an individual, but the distribution can be estimated by comparing age at diagnosis in screened and unscreened populations. However, apparently long lead times may be the result of the detection of cases which would resolve spontaneously. Earlier treatment is not inherently of benefit and should be shown to improve outcome without serious side effects compared to the treatment of clinically presenting cases.

In 1968, the World Health Organisation published Wilson and Jungner's guidelines (Table 2.1) for assessing whether a screening programme is appropriate.[6] Each of these criteria will be considered in relation to the screening programme for CDH.

Table 2.1 Wilson and Jungner's criteria for an appropriate screening programme

(1) The condition sought should be an important health problem.

(2) There should be an accepted treatment for patients with recognised disease.

(3) Facilities for diagnosis and treatment should be available.

(4) There should be a recognisable latent or early symptomatic stage.

(5) There should be a suitable test or examination.

(6) The test should be acceptable to the population.

(7) The natural history of the condition, including development from latent to declared disease, should be adequately understood.

(8) There should be an agreed policy on whom to treat as patients.

(9) The cost of case-finding (including diagnosis and treatment of patients diagnosed) should be economically balanced in relation to possible expenditure on medical care as a whole.

(10) Case-finding should be a continuing process and not a "once and for all" project.

2.2 BACKGROUND

2.2.1 Definition and terms

Congenital dislocation of the hip (CDH) has been defined as "a congenital deformation of the hip joint in which the head of the femur is (or may be) partially or completely displaced from the acetabulum. The term embraces secondary hip dysplasia whether or not hip instability or dislocation persists."[1] *Acetabular dysplasia* is used to describe a poorly formed and shallow acetabulum (socket in the bony pelvis). The term *luxated* may be used synonymously with *dislocated*, and *subluxated* with *partially dislocated.* Hips which can be induced to dislocate under provocation are considered *dislocatable* or *subluxatable*, although the latter term encompasses lesser degrees of displacement.

CDH as a disorder poses problems, as limited understanding of its natural history and the timing of the critical events leading to dislocation[7-9] has created uncertainty about how the condition should be defined.[10-13] At least fifteen different descriptors of CDH have been advocated or used in the literature.[10,11,13,14] It has long been assumed that CDH develops before birth, and is detectable at birth, but even with conscientious and expert screening, CDH has continued to present after the neonatal period. This, together with medico-legal considerations, has led to use of the adjective *developmental* rather than *congenital* (existing at birth). In addition, the term CDH also includes hips which, while not strictly dislocated at the onset of treatment, are assumed would progress to dislocation in the absence of treatment. The word *displacement* is therefore sometimes substituted for *dislocation.* Since the debate continues about whether a dysplastic acetabulum results in a dislocated hip, or whether the action of a displaced femoral head

produces a dysplastic acetabulum, the term *developmental dysplasia of the hip* may also be used.[12]

2.2.2 Aetiology

The aetiology of CDH remains unknown[15] but is considered the result of interacting genetic and environmental factors. The results of family studies,[16-18] which report an increased prevalence of CDH between siblings and parents of probands, indicate that there may be a genetic predisposition to CDH based on polygenic multifactorial inheritance. Twin studies have found the prevalence in the co-twins of dizygotic pairs to be no more than for siblings but in monozygotic pairs to be up to 34%.[19] Differences in prevalence between different ethnic groups have been reported and this cannot be wholly attributed to cultural differences in child care practices, e.g. the use of swaddling, although after a national campaign in Japan to avoid prolonged extension of the hips and knees of infants during the early postnatal period, the prevalence of CDH was reported to have fallen from 5-6% to less than 0.4%.[20]

Maternal hormones may induce joint laxity and facilitate hip displacement and have been thought to explain the consistently higher prevalence of CDH in female compared to male children.[19] However, this has not been directly proven and joint laxity may be familial.[21] Breech presentation at delivery or during the last trimester occurs more commonly in children with CDH than the population at large.[22] This, and the higher proportion of children with only a left hip affected, the side more commonly held adducted against the maternal sacrum[15] suggests that fetal positioning, particularly leg folding, affects the propensity of the hip to

dislocate. In some, although not all, studies, a higher prevalence has been reported among winter- and first- born children.[22]

2.2.3 History

Although it is said that more than 2,400 years ago Hippocrates believed CDH could be cured by early treatment,[23] it was not until 1847 that Dupuytren[24] gave the first modern account of CDH. He was the first to demonstrate the lack of development of the acetabulum, later referred to as dysplasia. Roser proposed a test for the early diagnosis of CDH in 1879,[25] which was supported by the work of Le Damany and Hilgenreiner.[26,27] However, it was Ortolani's paper of 1937[28] that was widely acknowledged and the clinical test for a reducibly dislocated hip bears his name. Nearly thirty years later, Barlow[29] proposed a modification to this test to allow the detection of dislocatable hips with the inference that these hips might later spontaneously dislocate and remain dislocated. This modification is now in general usage and is widely known as the Ortolani-Barlow (O-B) test.

Routine neonatal screening for CDH was introduced in Malmö, Sweden, in 1952 by von Rosen and his colleagues[30] who treated those children found to have neonatal hip instability (NHI) with abduction splinting. By 1958, they had reported only one false-negative result among 24,000 infants born in their obstetric unit since the start of screening. This child had required emergency care for an unrelated condition and had been referred to another hospital before the screening examination could be carried out. Although routine clinical screening for NHI was practised by some UK centres in the early 1960s,[31,32] it was not until 1969 that the Standing Medical Advisory Committee[33] formally recommended screening of

all neonates in England and Wales. These recommendations stipulated that examination for congenital dislocation of the hip should be carried out and recorded for every infant in the early neonatal period, using the O-B test, as described below, with a further examination before 3 months of age. In 1986, the Standing Medical Advisory Committee and the Standing Nursing and Midwifery Advisory Committee revised these recommendations and produced a more detailed report.[1]

2.2.4 Clinical screening examination

The Ortolani-Barlow test is carried out on each hip, either separately or concurrently, with the infant lying supine on a firm surface, the hips and knees flexed to 90°. The examination must be performed using warm hands with the infant relaxed, since the increased muscle tone around the hip joint of an angry crying baby may make an unstable hip joint appear stable. With the middle finger of each hand held over the greater trochanter, and the thumb over the lesser trochanter, an attempt is made to abduct the legs as far as possible (usually about 75°). The Ortolani procedure to detect a dislocated hip is performed by applying gentle upward pressure to the greater trochanter in an attempt to induce a potentially dislocated femoral head back into the acetabulum. Reduction is usually accompanied by a palpable clunk, which may or may not be audible. The Barlow modification to provoke dislocation in a located but a potentially unstable hip joint is carried out with the leg flexed and the hip initially in 45° of abduction. The hip is adducted and pressure applied to the lesser trochanter in a lateral direction while an attempt is made to provoke the femoral head gently out of the acetabulum and then reduce the dislocation. This is done by alternating pressure

over the lesser and greater trochanter with the thumb and middle finger respectively. In each part of this examination, the movement of a dislocated or dislocatable femoral head should be palpable but may not exceed 0.5cm, and may easily be missed by an inexperienced examiner. In larger infants, full abduction may be more difficult. This may be due to increased muscle tone, but may also reflect an irreducibly dislocated femoral head. The O-B manoeuvre may not detect an irreducibly dislocated femoral head[34] and thus leg length and thigh and buttock creases should be examined for asymmetry that may be indicative of CDH. Infants with bilateral irreducibly dislocated hips may therefore be particularly difficult to detect at birth.

2.2.5 Current UK Policy

Screening by the (O-B) test is currently advised on three occasions: within 24 hours of birth, at discharge from hospital of birth, and at 6 weeks of life (Table 2.2). It has been suggested that infants who test positive should not be repeatedly examined by a primary screener but referred for further assessment by a senior clinician. This is in view of the unproven suggestion that clinical examination may encourage an unstable hip to become frankly dislocated.[4,35] If diagnosis is confirmed, treatment with a splint appliance is usually initiated to maintain concentric reduction of the femoral head for a variable period until it is considered that stability of the hip has been achieved. The finding of a ligamentous click is not considered an indication for treatment[1,36,37] although this policy has been questioned and is controversial.[38-41] Some of this confusion may be due to inadequate translation of Ortolani's original term as a 'click' rather than as a 'sign of the jerk', as recently noted by Macnicol.[42] Additional clinical examinations for

'classic' signs (limited abduction, asymmetry of buttocks/thigh creases) and abnormal gait are also advised in later infancy and childhood, including at school entry. It is recommended that children at high risk of CDH should be regularly examined until they are at least at walking age. The factors which are considered to place children at high risk are: family history of the condition, infants presenting by the breech, other congenital postural deformities, birth by caesarean section, oligohydramnios and fetal growth retardation. In the latest edition of 'Health for All Children' in which the aim is to assemble current knowledge for a variety of childhood conditions and present guidelines for best practice, these recommendations are largely reiterated, but in addition, it is proposed that the examinations at 15-21 and 24 months should be replaced by a single examination at 18-24 months to coincide with other examinations.[43]

Table 2.2: Summary of current UK recommendations [reproduced from 1986 SMAC report][1]

		Examination for CDH		
	Age	**Ortolani-Barlow**	**Classic signs[1]**	**Gait**
a.	within 24 hours of birth	+	+	-
b.	on discharge from hospital	+	+	-
c.	6 weeks of age	+	+	-
d.	6 - 9 months	-	+	-
e.	15 - 21 months	-	+	+
f.	24 months +	-	+	+

[1] limited abduction, leg shortening, asymmetry of thighs, flattening of buttock,

2.3 SCREENING AND CDH

This section considers the current UK policy for screening for CDH, described above, in the light of the Wilson and Jungner criteria, as listed in Table 2.1.

2.3.1 Is CDH an important public health problem?

Sequelæ of untreated CDH

Established dislocation of the hip occurs when there is complete loss of normal contact between the acetabulum and the femoral head.[44] If left untreated, the limb on the affected side usually becomes shortened with a resultant limp[15] and thus in the absence of screening, CDH usually presents after walking age.[45] The extent to which the recognised complications - hip or low back pain, knee pain and

deformity, or degenerative changes in the hip joint - will develop in those with untreated hip dislocation is uncertain but they are considered more likely to occur in those with bilateral dislocations and in those who have developed a 'false' acetabulum between the femoral head and the ileum.[46] Published reports, based on small numbers of non-randomly selected cases of untreated CDH,[44] indicate that functional impairment is common and increases with age but is not inevitable.[15,46] Between 11 and 41% of those with untreated complete dislocation remained free of pain when followed for an average of 50 years,[15,46] and, in one 74 year old, hip dislocation was first diagnosed post mortem.[15] In addition, it has been estimated that 40% of cases of osteoarthritis are due to unrecognised and untreated hip dislocations.[47]

Individuals with subluxed hips tend to develop symptoms at an earlier age and are thought to have a worse prognosis than those with complete dislocation.[15] In one series, mean age of symptom onset with untreated subluxation was 37 years in women and 54 years in men, with radiological onset of degenerative joint disease usually occurring during the following decade.[15] In severe subluxation, hip pain may start as early as the second decade.[46]

Treatment of established CDH

The objective of treatment is to reduce or eliminate the pain, deformity, abnormal gait and premature osteoarthritis associated with untreated CDH. A wide range of surgical procedures has been developed to treat dislocated, subluxated or dysplastic hips[48,49] with the basic aim of achieving a stable and deep reduction of the femoral head into the true acetabulum.[50] These procedures are usually

classified as either 'closed' (manipulative reduction without direct surgery to the hip joint)[51,52] or 'open' (direct surgery to the hip joint).[53,54] Since surgical practice in the treatment of CDH has changed substantially in the last half century,[49,50] the prognosis for current treatment is likely to differ from that of the treatment available 30-40 years ago. Evaluation is further complicated by the lack of trial evidence and the small size of case series.

Long-term follow up to skeletal maturity and beyond, is required for an accurate assessment of the prognosis of treated dislocation, as a hip that is anatomically and radiologically poor may function well during childhood but deteriorate subsequently.[53,54] Radiological evidence of osteoarthritis was present in more than half of 264 subjects treated for CDH during the first half of this century who were successfully traced between 10 and 50 years following completion of treatment, when half were 20-40 years old and one fifth over 40.[51] Half of those with osteoarthritis had developed hip pain by the age of 40. Clinical and radiological outcome for those treated by closed reduction alone was assessed as good or better in 78% of unilateral dislocations and in 51% of bilateral dislocations. Outcome was worse for those bilateral dislocations first treated at 3 years and over, and only 28% were deemed good or better. In a more recent series of 42 children first treated between 1 and 3 years of age, 75% of whom had been treated with closed reduction, 71% of 51 treated hips were considered radiologically normal.[52] The prognosis for those treated by open procedures was initially less favourable than for closed procedures, but this may have reflected lesser severity and younger age at presentation in those selected for closed procedures.[50] In addition, with the subsequent advances in surgery, the prognosis for those treated with open surgery

has improved.[50,53-56]

While the quality of the initial reduction is considered an important prognostic factor,[57] subluxation, dysplasia and premature onset of degenerative joint disease have been reported to develop despite an apparently satisfactory initial reduction.[54,58] Overall, prognosis is considered better in those diagnosed and treated at a younger age because the potential to remodel the acetabulum may be lost in the older child,[48,50,58] although the possibility of avascular necrosis may be lower in an older child.[59] If treatment is not started until the end of the first year of life, the chance of the child having a functional pain-free hip may be reduced to about 75%.[53] Bilateral dislocations, which have a worse prognosis than unilateral dislocation, are usually diagnosed at a later age.[52,53]

Avascular (or ischaemic) necrosis is an iatrogenic and potentially serious complication of treatment where the blood supply to the femoral head is reduced and the normal development of the hip joint impeded. While in some series, up to two-thirds of treated hips have been affected[58,60], this proportion has fallen recently, possibly reflecting the adoption of the 'human' rather than the 'frog' position during post-reduction immobilisation[49,60] and the use of routine pre-operative skin traction.[50] In the earliest series,[55] only 21% of hips treated by open reduction were considered radiologically normal after 20 years and 41% had developed avascular necrosis. In those treated more recently,[50,56] radiological normality was observed in 51-78% after 20 years and avascular necrosis in 2-11%.

Thus, for many children, CDH results in functional and anatomical hip pathology,

despite multiple operations which entail prolonged hospital admission or immobilisation at home, frequently during a critical period of growth and development.

Prevalence

The *prevalence* of CDH is usually presented as the number of affected children, rather than number of affected hips, per 1000 live births. This is termed birth prevalence, the proportion of children affected, rather than *incidence*, the frequency with which new cases arise, because the denominator excludes still births, deaths and terminations of pregnancy.[61] Children with hip dislocation acquired secondary to another disorder, such as cerebral palsy, or spina bifida, or those with CDH as part of a syndrome, are generally excluded from the numerator.

Prevalence of CDH before screening was introduced

There are few data available regarding the prevalence of CDH in the UK before screening was introduced but the prevalence in unscreened populations, in which CDH is usually detected after walking age,[45] is known to vary greatly between different countries and different ethnic groups.[62] However, in his often-cited review of 1986[63] Leck reported that within Northern European populations, the prevalence of CDH ranged from 0.8 to 1.6 children per 1000 live births (midpoint estimate 1.2), and a figure of between 1 and 2 per 1000 is generally assumed.[64-66] This variation may reflect several factors, including differences in case definition over time, different methods of case ascertainment, real variation over time, or variation in the prevalence of familial, genetic[18,21] or environmental factors known to increase the risk of CDH.[67] The focus in this thesis will be on UK studies.

A further consideration is the precision of the studies cited. Although CDH is considered a relatively common malformation of childhood, a large population needs to be studied to estimate the prevalence with confidence. Most studies have been small in relation to the rarity of the condition and the estimates of prevalence sensitive to the identification of one or two additional cases, and vulnerable to migration. Incompleteness of case ascertainment may arise from the practical difficulties of ensuring all members of a birth cohort who develop CDH are identified, and not allowing sufficient time for all cases of a cohort to have presented. Premature publication of study findings may mean that some later-presenting children are omitted from published reports of prevalence. For example, 13 late presenting cases had not been ascertained at the time of the first publication of one Aberdeen series and prevalence, initially reported as 1.8 per 1000 live births,[68] was subsequently estimated to be 2.1 per 1000 live births.[69] Knowledge of the background prevalence is fundamental when evaluating a screening programme.

Leck's estimate of the prevalence of CDH in unscreened populations of northern European origin was based on 7 studies,[69-74] of which two were in the United Kingdom: one in Birmingham (Leck's own),[72] and one in Scotland.[69] The former, a multiple source retrospective study of children aged 5 years and under, reported a prevalence of 0.91 per 1000 live births, while the latter, an unpublished study cited in another publication,[69] estimated prevalence to be 1.5 per 1000 live births. Other studies[16,75,76] identified from the bibliographies of other publications but not included in Leck's review, have reported cohorts followed to the age of 3, 2 and 2 years respectively. From these studies, a prevalence of 0.66, 0.67 and 0.85 per

1000 live births has been reported in Birmingham,[16] Liverpool,[75] and Wales[76] respectively. It would appear from these and subsequent studies[68,69,77] that the prevalence in Scotland is unusually high and in England and Wales lower, ranging from 0.66 to 0.91 per 1000 live births.

2.3.2 Is there a recognisable latent or early symptomatic phase of CDH?

To be suitable for prevention by screening, a disease must pass through a preclinical stage during which it is undiagnosed clinically but may be detectable. The preclinical phase begins when the pathologic process is first present, which in the case of CDH, is believed to be before birth. In the absence of screening, the preclinical phase ends when the affected person seeks medical attention, which is likely to be when a child with CDH is discovered to limp or waddle on walking, and is usually in early childhood. The O-B test is not, strictly speaking, a test for CDH but rather for neonatal hip instability which is thought to be the preclinical stage of CDH.

Prevalence of NHI

Hips with NHI can be classified as Ortolani-positive or Barlow-positive. The prevalence of each has been reported only since the 1940s and the 1960s respectively, but in practice, distinction is not usually made between the two. The prevalence of NHI in northern European populations is reported to range from about 2.5 to 20 children per 1000 live births[63]. Some difficulties of complete case ascertainment associated with later-presenting CDH do not apply to neonatal cases, but interobserver variation may be much greater, as discussed below. In addition, authors may differ in their criteria for abnormal hips. In one centre, the

prevalence of NHI was reported as 2.5 per 1000 live births, but an additional 9.2 per 1000 live births were reported to have instability in which the femoral head could be moved to the rim of the acetabulum but not further.[78] Some variation may be attributed to the age at examination, since most neonatal instability is known to resolve spontaneously (without treatment) shortly after birth.[29,38,79] These children are not always treated, although it has been suggested that such infants are at risk of subsequent dislocation and should therefore be treated.[29,45,79]

2.3.3 Is there a suitable test?

The incorrect designation of infants as test-positive or test-negative may stem from either the test itself or its interpretation. For example, the O-B test may not reveal latent hip instability, or the screener may decide incorrectly that a child requires treatment based on an equivocal test finding. Without a confirmatory diagnostic test, the sensitivity and specificity of the O-B test cannot be directly calculated but the specificity, at least, is likely to be poor since NHI affects between 3 and 20 children per 1000 live births while the prevalence of CDH before screening was introduced ranged from 0.66 to 1.5 children per 1000 live births. Similarly, the optimum timing of the clinical examination when the test is most sensitive and specific, is not clear. However, testing needs to be undertaken at a convenient time for both parents and medical staff to limit costs, and to maintain a high coverage. Most children in the UK are currently born in hospital and constitute a 'captive' population for a few hours. Hence coverage for the first initial O-B test is thought to be high.[80] Although it may cause some discomfort, examination lasts only approximately 1.5 minutes.[81]

It has been suggested that poor test performance may reflect lack of expertise on the part of the primary screener,[42,82-89] although cases have been missed by expert examiners.[38,90] A poor level of agreement between different examiners was reported from one study[91] in which hip instability was independently diagnosed by a paediatric orthopaedic surgeon and a neonatal paediatrician in 51 of 12 891 newborn infants examined, but was not detected by one of the examiners in a further 25. Another 28 infants, judged by both examiners to have stable hips at birth, were subsequently referred for orthopaedic treatment. There is limited consensus among experts as to what constitutes an acceptable clinical examination.[92] In one study, only 40% of those responsible for hip screening could correctly identify both the dislocated and dislocatable hips of a hip simulator model.[92] The need for formal training and expert supervision of those responsible for primary clinical screening has been emphasised.[62,89,93] In one study, screening performance was better when undertaken by trained senior physiotherapists supervised by orthopaedic surgeons than by junior paediatricians.[84]

Risk factors for NHI

The term *risk factor* has been defined[94] as "an aspect of personal behaviour or lifestyle, an environmental exposure, or an inborn or inherited characteristic, which on the basis of epidemiological evidence is known to be associated with a health-related condition". It need not be a causal factor. Determining risk status itself constitutes an additional screening test.[3]

NHI shares many of the risk factors associated with CDH. These risk factors include pregnancy characteristics such as oligohydramnios, intrauterine growth

retardation and breech presentation; congenital abnormalities affecting the skeletal and neuromuscular system; a positive family history of CDH; and being firstborn or a girl.[1,45,63,79] Genetic factors and differences in postnatal infant swaddling practices are also thought to contribute to the observed variations in prevalence.[20,63] The effectiveness of selective programmes of follow up for children at high risk depends on the predictive ability of the risk factors chosen and on the ease of eliciting the risk factor information. For example, gender and mode of delivery are evident at birth, but a positive family history may be missed. However, although CDH occurs several times more frequently in girls than boys, gender is not sufficiently predictive of CDH to be a useful risk factor. Similarly, CDH is found more often in White and first born children but these characteristics are too common to be of practical value as risk factors in a screening programme.

Continued diagnosis of clinically presenting CDH

A negative impact of screening on the continued diagnosis of clinically presenting CDH has been reported which has been attributed to a reduced index of clinical suspicion.[95,96] Although increased vigilance for dislocated hips during later infancy and around walking age has been advocated,[65,95] the implications for training of those responsible for child health surveillance have not been addressed. The Hall report discusses the key role of parents in the detection of abnormalities and stresses that some conditions require a special search by health professionals.[43] CDH falls into this category.[80]

2.3.4 Is early conservative treatment effective?

Screening may present an opportunity for improving outcome of CDH through

earlier diagnosis that facilitates earlier conservative treatment. Conservative treatment is undertaken with the expectation that the trauma and potential complications of invasive surgery will be avoided and outcome be improved. It is usually undertaken on an outpatient basis.

A variety of plastic splints may be used for early conservative treatment, but each is designed to hold the infant's hips flexed in abduction, to encourage concentric reduction of the femoral head within the acetabulum.[30,97] Except for one trial in Thailand[98] in which a plaster of Paris hip spica was compared to a plastic splint appliance, the performance of the various splints has not been assessed in clinical trials, but only in a few 'before and after' studies of outcome. Criteria for failure may vary[99], and both the rate of failure and rate of complications need to be considered. In addition, full reduction by a splint may not be possible in some children[100], in which case conservative treatment, if attempted, may be harmful.

Published studies comparing different appliances have found in favour of the von Rosen splint over the Frejka pillow,[99,101] but had no preference for the Pavlik harness over the Frejka pillow.[102] In one paper, the authors recommended the use of the cheaper Becker abduction pillow in preference to the more expensive Pavlik harness.[103] Double nappies, used instead of formal splinting, have been found ineffective.[95,104] In the trial of the hip spica and a plastic splint, in which most children were followed up to at least two years of age, the two methods were to be equally effective[98] but the plastic splint was recommended for its lightness, and because it can be removed for bathing the child.

The infant hip is sensitive to ischaemia resulting from undue pressure. Avascular necrosis of the femoral head has been reported to affect up to 28% of the hips of infants treated with a splint appliance[83,105-109] and has also been documented in the contralateral normal hip.[110] Other hazards of splinting, such as pressure sores, tibial torsion and femoral nerve palsy have been reported.[109,111,112] The implications of even a short period of splinting for motor, social and psychological development are not known.[113] Evidence from a British-based support group for parents of infants with lower limb abnormalities, including CDH,[114] indicates that for some families at least, the psychological, social and financial sequelae of hip instability may be considerable. Poor parental adherence is cited as a factor in unsuccessful splinting[115] and although the extent of this is unknown, has lead some authors to propose the use of nonremovable splints.[101] However, the Frejka pillow, 'frog' casts and others which enforce flexion and full 90° abduction are thought to have higher rates of iatrogenic complications.[116,117] The Pavlik harness, which is a removable non-rigid splint, is associated with a lower rate of complications[105] attributed to the greater degree of mobility it allows within a controlled range.[112,118] The age at which conservative treatment becomes ineffective or is no longer feasible is not clear, although in one study, outcome did not differ between those initially treated at birth and those initially treated at up to 5 months of age.[119]

Difficulties in deciding whether to treat NHI arise because there is a spectrum between a "normal" hip in which the femoral head is located firmly within the acetabulum and a frankly dislocated hip where the femoral head lies at rest wholly outside the acetabulum. This dilemma is the direct result of screening which has led to the identification of NHI and dysplasia (the latter since the advent of

ultrasound), which are assumed to be preclinical stages of CDH. However, the association between early hip instability and acetabular dysplasia is unclear,[11,66] as is the relation of either of these conditions to the later development of established hip dislocation.

In *screened* populations, the prevalence of neonatal splinting is anywhere from 3 to 30 times more common than the prevalence of established dislocation in *unscreened* populations which represents a positive predictive value of, at best, 34%. Thus, relatively large numbers of infants may be receiving unnecessary treatment as a result of the current screening policy, with the consequent restrictions and potential complications associated with splinting. An adverse psychological impact on families given a false positive screening result in their newborn child has been reported in other neonatal screening programmes,[120] but there seem to be no similar studies in relation to screening for CDH.

2.3.5 How effective is the screening programme for CDH?

Measuring outcome

The main objective of the CDH screening programme is to ensure the normal development and function of the hip at the end of the period of childhood and adolescent growth. Radiological evaluation of the hip in early childhood to assess acetabular angle and presence of avascular necrosis has been advocated as an earlier and more appropriate measure of outcome,[121-124] but it has not been widely embraced[7,25,53,84,89,100] possibly since some authors doubt that adult function can be predicted before skeletal maturity, perhaps not even until the third decade.[125] The importance of correct positioning of the hip on X-ray is emphasised.[126]

A national evaluation of the current screening programme for CDH has not been attempted but several interested centres have undertaken local evaluations. These studies are all retrospective or prospective cohort studies which use surgery for partial or complete hip dislocation as a measure of outcome.[69,84,127] The figures are vulnerable to the ascertainment issues discussed above (section 2.3.1) and to the varying definitions of a 'late' case of CDH, which might, for example, include all children detected after the initial screening examination at birth or only those detected after 2 months of age. It is important to distinguish between surgery required following late detection of CDH (the false negatives of the screening programme)[63,68] and that required following the unsuccessful conservative treatment of a screen-detected hip abnormality.

A summary of the published reports of the prevalence of surgery for CDH in the UK, identified through searches of the MEDLINE and BIDS computer databases and from the bibliographies of published papers, is provided in the evidence table below (table 2.3), but detailed information on study methodology could not always be extracted, in particular the length of time after birth for which cases were sought. Section A of table 2.3 includes studies in which an author-defined prevalence of late CDH was reported, and section B, studies in which the total prevalence of surgery (including children who received prior abduction splinting) was reported.

Table 2.3 Prevalence of a first operative procedure for CDH

publication year	first author	place	period	prevalence 10^{-3} (to 2dp)	Size
A: Prevalence of a first operative procedure for author-defined late CDH					
1962	[29]Barlow	Salford	1957-60	0	3647
1963	[90]Barlow	Salford	1957-	0.10	9680
1964	[31]Barlow	Salford	- 1964	0.07	14960
1967	[32]Finlay	Uxbridge	1962-66	0.07	14594
1972	[104]McKenzie	Aberdeen	1960-69	0.91	76675
1972	[128]Mitchell	Edinburgh	1962-68	0.12	31961
1972	[34]Wilkinson	Southampton	1965-68	1.28	6272
1972	[129]Williamson	Northern Ireland	1960-70	0.56	337700
1977	[130]Jones	Norwich	1968-72	0.58	29366
1978	[89]Noble	Newcastle	1964-74	0.15	25921
1978	[131]Place	Leeds	1974-76	0.78	26908
1980	[132]Galasko	Salford	1975-78	0.83	11980
1981	[133]Dunn	Essex	1972-76	0.81	36918
1981	[69]McKenzie	Aberdeen	1970-79	1.11	53033
1982	[134]Bertol	Edinburgh	1969-78	0.62	44953
1982	[95]Catford	Southampton	1965-78	0.87	76724
1984	[40]Cunningham	Nottingham	1979-81	0.89	7864
1985	[83]Dunn	Bristol	1970-79	0.44	23002
1985	[83]Dunn	Bristol	1970-79	0.88	103431
1987	[36]Bernard	Solihull	1977-83	0.71	21004
1987	[135]Dwyer	Birmingham	1976-86	0.07	28000
1988	[136]McKibbin	Wales	1981-84	0.58	15561
1989	[137]Clarke	Coventry	1977-86	0.77	44073
1989	[138]Kernohan	Northern Ireland	1976-85	1.00	271240
1990	[139]Jones	Swansea	1987	0	3789
1990	[139]Jones	Swansea	1979-85	2.20	n/a
1990	[42]Macnicol	Edinburgh	1962-86	0.50	117256
1990	[140]Myles	Peterborough	1980s	2.18	3205
1990	[140]Myles	Peterborough	1980s	0.73	5456
1992	[84]Krikler	Birmingham	1980-90	0.08	37511

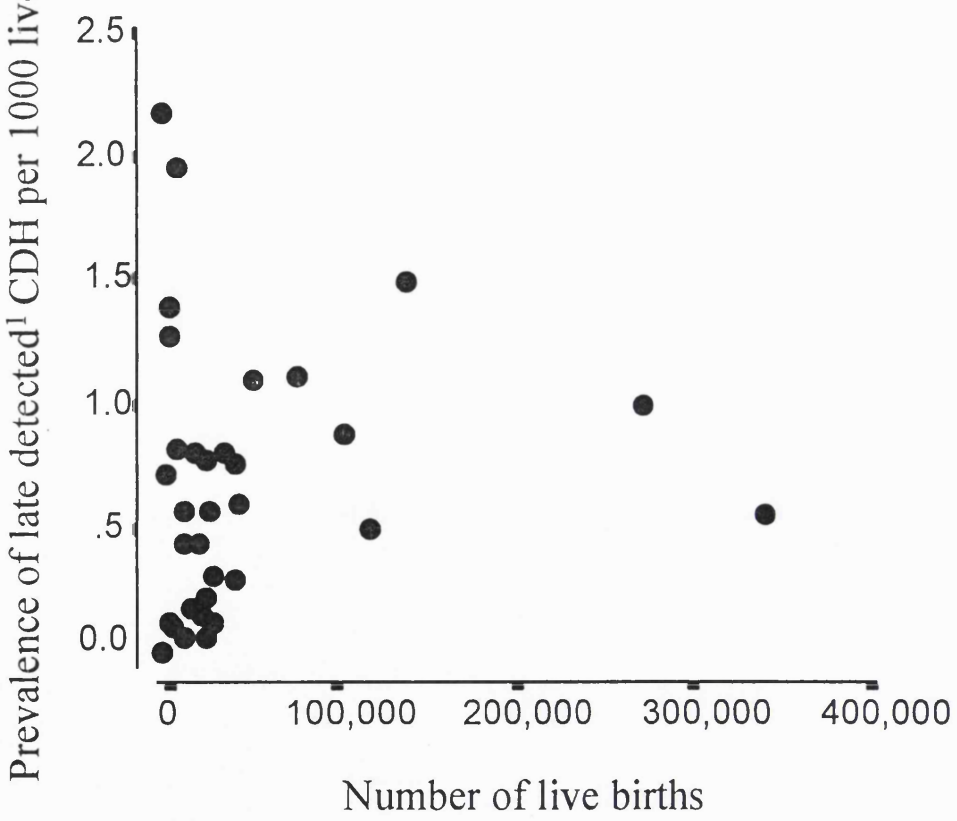
publication year	first author	place	period	prevalence 10^{-3} (to 2dp)	Size
1992	[84]Krikler	Birmingham	1980-90	0.32	31350
1993	[68]Lennox	Aberdeen	1980-89	1.30	67093
1994	[141]Boeree	Southampton	1988-92	0.22	26952
1994	[86]Fiddian	Poole	1982-92	0.31	42421
1994	[86]Fiddian	Poole	1979-82	1.96	11251
1995	[142]Patterson	Northern Ireland	1983-87	1.65	138600
1995	[143]Vedantam	Sheffield	1989-91	0.13	7827

B: Prevalence of a first operative procedure for CDH after a late diagnosis or the failure of conservative treatment

1972	[104]McKenzie	Aberdeen	1960-69	1.12	76675
1972	[128]Mitchell	Edinburgh	1962-68	0.19	31961
1978	[89]Noble	Newcastle	1964-74	0.46	25921
1981	[69]McKenzie	Aberdeen	1970-79	1.83	53033
1982	[134]Bertol	Edinburgh	1969-78	0.82	44953
1982	[95]Catford	Southampton	1965-78	1.28	76724
1984	[40]Cunningham	Nottingham	1979-81	2.80	7864
1985	[83]Dunn	Bristol	1970-79	0.65	23002
1987	[36]Bernard	Solihull	1977-83	1.19	21004
1987	[62]Knox	Birmingham	1974-83	0.67	14426
1988	[136]McKibbin	Wales	1981-84	1.09	15561
1992	[84]Krikler	Birmingham	1980-90	0.45	37511
1992	[84]Krikler	Birmingham	1980-90	0.48	31350
1993	[68]Lennox	Aberdeen	1960-69	1.10	76675
1993	[68]Lennox	Aberdeen	1980-89	2.10	67093
1994	[141]Boeree	Southampton	1988-92	0.41	26952
1994	[86]Fiddian	Poole	1982-92	0.47	42421
1994	[77]SNAP	Scotland	1986-91	1.10	388182
1995	[143]Vedantam	Sheffield	1989-91	0.51	7827

Based on these studies, the mean prevalence (95% confidence interval) of author-defined 'late' CDH was 0.70 per 1000 live births (0.51, 0.89), but these data are positively skewed. The median (95% confidence interval), which may be a more appropriate summary, was 0.62 per 1000 live births (0.50, 0.83). The confidence intervals are wide but the confidence interval for the prevalence of a rare condition is wide even when a relatively large single population is studied. An alternative approach to obtain a summary estimate takes account of the differing samples sizes. In Table 2.3, 8 of the 36 studies the studies cited in section A were based on fewer than 10 000 children. Since a larger study may follow a cohort born over a longer period than a smaller study, case ascertainment may be improved because the study is ongoing when the children born at the start of the study present with late CDH. Conversely, publication bias and the less manageable follow up of large numbers of children may result in lower ascertainment for larger studies.[144] No trend in the prevalence of late-presenting cases with increasing study size is clear in Figure 2.2. However, larger study sizes improve the precision of an estimate. Assigning weights to each study corresponding to the reciprocal of an individual study's variance produced a weighted average (95% CI) of the overall estimate of the prevalence of late detected CDH of 0.88 per 1000 live births (0.85, 0.91). This suggests that larger studies tended to report a higher prevalence of late CDH. These data are, however, extremely heterogeneous ($\chi^2 = 400$, $p < 0.001$) and thus any pooled estimate should be viewed with caution.

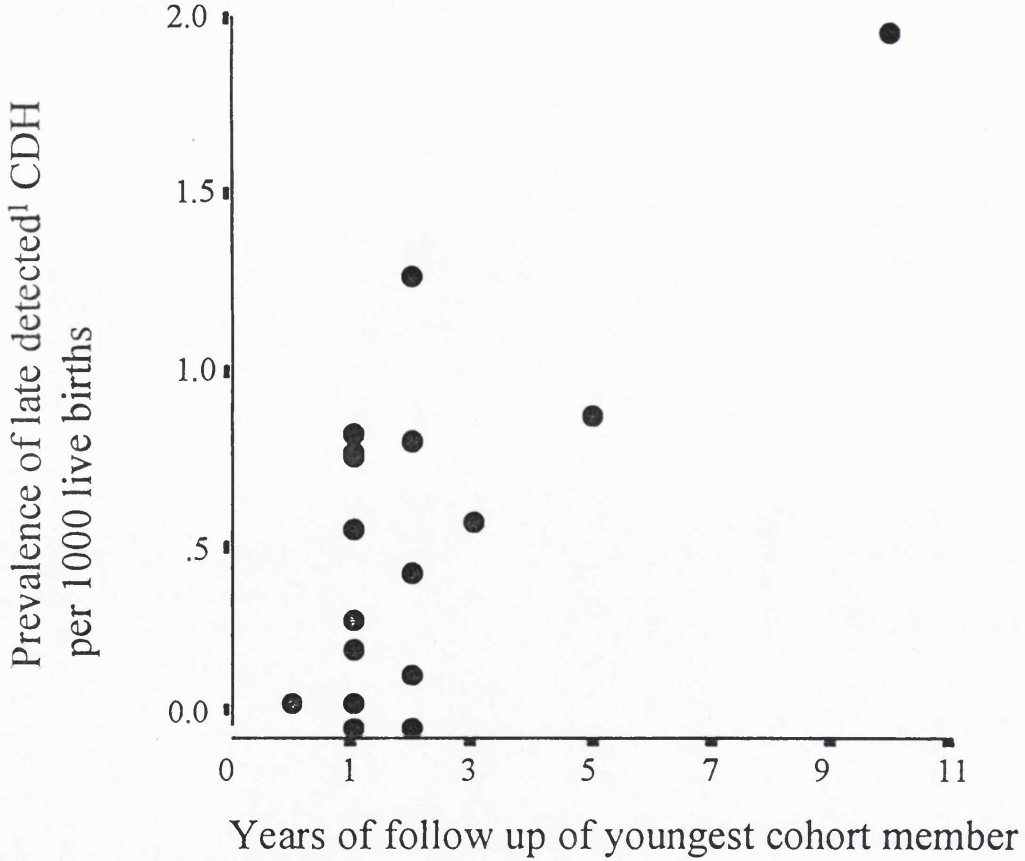
Figure 2.2 Association between study size and estimates of the prevalence of late detected [1] CDH, UK



[1]not detected by screening

A low prevalence of late detected CDH may indicate a more successful screening programme but there is evidence to suggest that variation in the completeness of case ascertainment may explain the reported differences. For example, a higher prevalence of surgery was reported from centres in which cohorts were followed for longer periods, (Figure 2.3).

Figure 2.3 Prevalence of late detected[1] CDH and length of follow up of cohort



[1]not detected by screening

Natural history

The natural history of CDH is not fully understood. Cases of CDH have continued to present after the neonatal period despite expert and conscientious screening. These false negatives may include cases not preceded by instability by 6 weeks of age, reflecting variability in the natural history of CDH.[7,38,145] Onset of CDH may not be from birth but from weight-bearing age, or some children who were not detected by screening may have had irreducibly dislocated hips which may not be detectable on the O-B test. Conversely, NHI is found in far greater numbers of children than is CDH.

Failures of early treatment

Based on the studies included in Table 2.3, section B, the median crude prevalence of surgery for CDH including children in whom early treatment did not resolve the abnormality was 0.82 per 1000 live births (95% CI: 0.48, 1.1), generally higher than the prevalence of late diagnoses and similar to the prevalence of surgery before screening was introduced. Leck reported the prevalence of surgery in children previously treated with abduction splinting to range from 0.11 to 0.25 per 1000 live births.[63] This raises questions about the efficacy of abduction splinting treatment. Surgery may be needed in up to 5% of infants treated with a splint appliance,[68,69,83,84,86,89,105,119,141,146] and higher figures have been reported.[127] This may reflect several factors including severity at presentation (particularly the finding, at diagnosis, of an irreducibly dislocated femoral head), failure to ensure concentric reduction during treatment, and

premature removal of the splint appliance by parents.[104,105,115] However, case ascertainment is likely to be higher in children already under medical supervision and those children who have been previously splinted may contribute proportionally less to the true crude prevalence of surgery than it appears from these figures.

In the case of the screening programme for CDH, it is not possible to calculate the detection rate or the false-positive rate directly because there is no confirmatory diagnostic test and treatment is usually started before the disorder becomes manifest. Inferences are drawn from the prevalence of abduction splinting and the prevalence of surgery. One method for calculating the detection rate and odds of being affected given a positive result (OAPR) has been described which uses a midpoint estimate of the prevalence of CDH in unscreened infants of 1.2 per 1000.[147] On this basis, data from two British centres[86,141] (both of which used ultrasound to examine clinically detected hip instability and to assess further high risk children) indicates a detection rate of 83% and an OAPR of 1:5.2 and 1:3.5, compared to 64% and 1:24 respectively from a centre using clinical screening alone.[83] However, these centres differ not only in relation to the use of ultrasound, but also in the personnel used to screen and the timing of diagnostic confirmation, which limits the extent to which inferences about the effectiveness of the respective policies may be made. These figures are subject to the limitations of cohort studies to evaluate screening for CDH discussed earlier (section 2.3.1) and assume a background prevalence of CDH of 1.2 per 1000, which may be an overestimate for

English and Welsh populations but an underestimate for Scottish populations (section 2.3.1). This method cannot be applied where a centre reports a prevalence of surgery that is higher than the estimated background prevalence of CDH, and it is clear that these data must be interpreted with caution.

Economics

A successful screening programme requires that the resources expended in the identification and earlier treatment of a condition are less than the resources averted and benefits gained. Since screening is conducted on a large scale, financial costs may be high even if the cost per individual screened is low.[148] The marginal cost of clinical screening for CDH as part of the routine neonatal examination is likely to be low. However, the average cost of treating a child with late CDH was calculated as Can$13,700 in British Columbia in 1984,[149] as GB£6,674 in Northern Ireland in 1991,[150] and most recently, as US$6,977.6 in Norway in 1993.[151] Costs will vary over time with changing treatment and trends to earlier discharge home, but the current cost to the health service of treating a child with late detected CDH is likely to be more than £4000, even before potential negligence claims.

There has been no trial of clinical screening but a decision analysis[149] found clinical screening to be cost-effective compared to no screening until the false negative rate reached 0.8 per 1000 live births.

2.4 ULTRASOUND EXAMINATION OF THE NEWBORN HIP

The perceived failures of the current screening policy, together with the implementation of universal primary ultrasound screening in some European countries,[152,153] have generated interest in the use of ultrasound as a primary screening test in the UK.[11,146,154] Selective primary ultrasound screening of infants at high risk of CDH (for example, those with a positive family history or breech presentation) has been proposed, but experience in the UK has not so far confirmed the success of this approach in reducing the prevalence of late cases.[137,139] Ultrasound may also have a role in reducing the number of false positives associated with clinical screening[154,155] and is, in addition, used in some centres to confirm concentric reduction of the femoral head with the splint appliance *in situ*, and to determine treatment duration.[137,146]

Screening with ultrasound may be with static or dynamic imaging. Static ultrasound imaging assesses morphology of the hip joint: specifically, the depth of the cartilaginous acetabulum and the location of the femoral head at rest.[153] With dynamic imaging, the morphology of the hip is examined with ultrasound while the hip is moved.[8,156] The two techniques may be used separately or in combination.[8,157] While much effort has been directed at grading the features observed from static imaging, the natural history and outcome of these grades have not been established.[8,9,158] It is also unclear whether neonatal acetabular dysplasia leads to CDH or is secondary to NHI. Although degenerative joint disease with onset in early adult life is frequently associated with acetabular dysplasia,[15] in a longitudinal study of untreated congenital hip disease in Navajo Indians,[159] spontaneous resolution of radiologically ascertained dysplasia was

observed in 10 of 14 children.

Clinical screening based on the O-B test identifies hip instability rather than dysplasia.[1,29] Some studies have compared clinical to ultrasound assessment,[8,9,141,146,152,160,161] but the level of agreement between tests that detect differing 'pre-clinical' phases is of less interest than their respective or combined predictive value for established dislocation. As with the O-B test, assessment of test performance is limited by the absence of a 'gold standard'. There may be wide variation in the level of agreement achieved even when standardised procedures for performing, measuring or reporting either static or dynamic hip ultrasound images are employed by well-trained examiners.[162,163] In the UK, there is currently no nationally agreed system for training and accreditation in neonatal hip ultrasonography, in contrast to Europe and North America.[156]

There has been one non-randomised trial of universal primary ultrasound screening in Norway[81] which showed that the addition of a dynamic and static ultrasound examination at 24-48 hours of life to universal clinical screening almost doubled the percentage of infants treated from 1.8 to 3.4%. In addition, 13% of all infants examined by ultrasound required follow-up for ambiguous findings compared to none for clinical screening alone. Ultrasound screening did not appear to reduce significantly the prevalence of false negatives, but the small numbers and a short period of follow up limit interpretation. A high proportion of ultrasound abnormalities identified by 48 hours of life have been shown to revert to normal by 12 weeks of age[164] and this, combined with the substantial increases in treatment and follow up reported following ultrasound screening from

Norway and Germany,[81,152] raises questions about the role and appropriate timing of this procedure. Universal ultrasound screening at birth and again at 4-5 weeks of age has been formally adopted in Germany.[11,165] The cost effectiveness of primary screening based on clinical or ultrasound examination, or some combination of both, was assessed as part of the Norwegian trial.[81,151] It was suggested that the most cost-effective approach was a strategy of screening all girls, but only those boys with NHI, breech presentation or a positive family history. However, another cost-effectiveness study, which used decision analysis[166] found neither universal nor selective ultrasound screening to be cost-effective. Neonatal acetabular dysplasia without hip instability has been characterised only since the 1980s with the advent of ultrasound and the study of large numbers of children will be needed to establish natural history.

2.5 CONCLUSION

Wilson and Jungner published their criteria for a screening programme in the year before the introduction of routine screening for CDH in the UK. The controversies which surround clinical screening for CDH attest to the problems created by introducing this programme before such recognised criteria for screening were met.[6,11,66,167] Some centres have reported a reduction,[79,83] some no change,[131,168] and others an increase[38,95,127,142] in the number of late presenting cases occurring since clinical screening was introduced. The effectiveness of clinical screening in altering the prognosis for those with hip instability is equally controversial, with some suggesting that, despite early treatment, there has been little impact on the numbers of children requiring at least one surgical procedure,[127] and others that it allows surgical treatment, if required, to be started

earlier and to be less invasive.[83] The effectiveness of clinical screening has been questioned for at least a decade, but there has been little progress in establishing whether it is worthwhile. Evaluation of the current policy is complicated by a number of factors which relate not only to the disorder and to the screening test but also to its management and implementation within the current UK programme. Centres with a special interest in managing CDH have reported their results, but a national picture of the performance of the screening programme is lacking.

While primary ultrasound screening has the potential to reduce the false positives and negatives associated with clinical screening, enthusiasm for this new technology has been tempered by the subsequent large increases in treatment and follow up reported. It was against this background that, in 1989, the Medical Research Council was asked by the Department of Health to establish a multidisciplinary Working Party (Appendix 2.1) to review existing policies for the screening and subsequent management of suspected CDH, with particular emphasis on the potential role of ultrasound imaging. The methods and findings of two separate but related national studies undertaken in the first part of this review are reported in this thesis. The first was a national survey carried out to identify current UK screening and management practices for CDH and to ascertain the potential for future trials. The second was a national surveillance study to obtain a nationally representative prevalence of treatment for CDH and NHI. The methodology for each study is described in the following chapter, and the results in Chapter 4.

3: METHODS

To identify current UK screening and management practices for CDH and to ascertain the potential for future trials.

3.1 NATIONAL SURVEY OF SCREENING AND MANAGEMENT

3.1.1 Survey respondents

In the absence of unequivocal and comprehensive recommendations for the screening and management of neonatal hip instability, and with little expert consensus,[11] considerable between-centre variation in screening practices was expected. All maternity units in the UK and Republic of Ireland were therefore included in the survey population.

Maternity units, and for each unit, a paediatrician responsible for the routine neonatal care of infants, were identified from the Directory of Emergency and Special Care Baby Units (SCBUs),[169] supplemented by a list of SCBUs in England and Wales compiled by the Neonatal Nurses' Association, the Medical Directory, the Health Services Year Book,[170] and a list from the Royal College of Obstetricians and Gynaecologists. The SCBU list provided the basis of the sampling frame because it was available on computer disk, included regional identifiers and named paediatricians. Since the target survey population comprised all maternity units, respondents were asked to list all units for which they provided neonatal cover. An appropriate respondent was identified by telephoning a unit if no name was listed.

In December 1993, a single named consultant paediatrician in each maternity unit was sent a postal questionnaire (Appendix 3.1) with a covering letter (Appendix 3.2) and reply-paid envelope, requesting information on the unit's current practices for the screening and management of CDH. Non-respondents were sent up to three reminders at seven week intervals, with further copies of the questionnaires in case the originals were not to hand. All secretaries of non-respondents were telephoned to ensure that addresses were correct and that questionnaires had been sent to the most appropriate paediatrician. The information departments of hospitals for which a questionnaire was *not* returned were telephoned, to confirm that they had maternity beds and to ascertain the annual number of births.

3.1.2 Questionnaire design

In the absence of clear evidence regarding best practice, this study was not intended to constitute an audit, but the current guidelines were used as a basis for the formulation of questions. Although the SMAC report of 1986 was written to clarify policy primarily for England and Wales, its recommendations were similarly implemented by Scotland, Northern Ireland and the Republic of Ireland.

A 'survey' has been defined as 'a technique of data collection, that is the systematic and structured questioning, either by interview or by questionnaire, of a relatively large number of respondents'.[171] Its design depends upon the question(s) a study is intended to answer. Although the

response to surveys between groups of professionals has been shown to exhibit wider variation than the response by patients and the general population,[172] greater familiarity with technical terms can be assumed. In one study, shortening a questionnaire improved response among general practitioners.[173] Care was taken to keep questions in this survey to a minimum.

The front of the four page questionnaire clearly identified the study, the researchers and research institution, and assured confidentiality. Respondents were asked to confirm whether they had been a consultant responsible for neonatal care in the past year, to list all maternity units for which they provided neonatal cover, the approximate number of live births in each unit for 1992 and to confirm whether the practices described applied to all units for which they were responsible.

Each subsequent page related to a different aspect of the screening programme to facilitate completion of the questionnaire, and in particular, to allow a respondent to skip easily the questions not applicable to their unit. The second page related to the organisation of routine clinical screening for neonatal hip instability, the third to screening by ultrasound imaging, the fourth to the management of infants at high risk of CDH and the remainder to the management of infants with clinically detected hip instability.

Tick boxes were used primarily, and open-ended questions kept to a

minimum. Filtered questions were indented and consistent sub lettering used. The last page provided space for additional comments, thanked the respondent for completing the questionnaire, and requested its return. The study address was included in case the reply-paid envelope was lost, and to aid the questionnaire's return when a paediatrician passed the questionnaire to another specialist for completion.

Details of second and third hospitals covered by a single respondent, in which practices differed from the first hospital, were requested from the respondent or other nominated paediatrician on an additional questionnaire unless details of the other units had been obtained independently. The responding paediatrician was asked to nominate a person responsible for the hip ultrasound service and a person responsible for the management of infants with dislocated or dislocatable hips who could be contacted for further details if necessary.

The questionnaires were coded on return and confirmed by the study supervisor (Dr Dezateux). Differences in coding were unusual and, when necessary, were clarified by further contact with the reporting clinician. The values of each variable were numbered and consistency of numbering was maintained between variables, for example, the code of a consultant paediatrician was the same for all questions. The replies to the questions regarding the use of ultrasound (question 8a) and the use of double nappies (question 21b) were given a summary code. All data were entered and validated with a double-entry system (Epi-info v.6, Atlanta).

3.1.3 Pretesting the questionnaire

A copy of the draft questionnaire was sent to 21 paediatricians across the UK in a variety of NHS settings, 2 paediatric orthopaedic surgeons, 2 paediatric epidemiologists, and 2 radiologists, and amended in the light of their comments. The main changes were to separate the questions for dislocatable and dislocated hips, to request the usual duration of splinting and to ask whether X-ray or ultrasound was used to monitor progress. Concerns about children who are discharged before a second neonatal examination can be undertaken were addressed by including a question relating to the proportion of children who received a second examination. The categories for the reporting of ultrasound were clarified. Careful reading by some piloters introduced rephrasing into some questions, for example question 20 was changed from 'Do you arrange further assessment...' which may have been ambiguous to 'Is further assessment arranged...'.

3.2 SURVEILLANCE STUDY

To estimate a nationally representative prevalence of treatment for CDH and NHI.

3.2.1 Introduction

As discussed in Chapter 2, the reported prevalence of CDH in unscreened populations in the UK ranges from 0.66 to 1.5 per 1000 live births while the median prevalence of CDH detected after the neonatal period in screened populations is 0.62 per 1000 live births, and the prevalence of abduction splinting for NHI ranges from 2.5 to 20 per 1000 live births. The variable success of the implementation of the screening programme locally is not readily explained, and is likely to be complicated by the heterogeneity of case definitions, periods of follow up and methods of case ascertainment. A national picture was required.

3.2.2 Routine data sources

The OPCS National Congenital Anomaly System (formerly the OPCS Monitoring Scheme for Congenital Malformations)[174] was established in 1964 to monitor congenital malformations in England and Wales but included until 1995 only those abnormalities detected at or within ten (formerly seven) days of birth. Reporting is voluntary and is known to be incomplete for CDH, particularly for later-presenting cases.[175] The prevalence of CDH calculated from notifications to OPCS was 0.29 per 1000 live births in 1992,[176] which indicates substantial under ascertainment of cases. A comparison to the Liverpool Congenital Malformations Registry found 35.5% of the malformed children reported to the latter

within 7 days of birth were not notified to OPCS.[177] Conversely, presumptive positive cases are reported to the OPCS scheme and diagnosis may not be confirmed. Similarly, the Scottish Morbidity Record, SMR11, which is completed for every Scottish infant discharged or transferred in the neonatal period, has been shown to contain a high proportion (78%) of incorrect diagnoses of congenital anomalies, despite identifying only 28% of all confirmed cases.[178]

Hospital Episode System (HES) data are collected for administrative purposes as a measure of hospital in-patient activity. One episode comprises a single period in hospital within a specialty. Its reliability as a means of identification of cases has been questioned[179] and it has been recommended that cases thus identified should be confirmed by reference to the original medical records.[180] The expense of this on a national basis precluded the use of national HES data for this study. Furthermore, since HES documents in-patient episodes, periods of abduction splinting, usually managed on an out-patient basis are not included.

As appropriate routine national sources of data for CDH were lacking,[88] cases were ascertained through active reporting by consultant paediatricians and consultant orthopaedic surgeons.

3.2.3 History of surveillance

The term *surveillance* was used until around 1950 to describe the activity of monitoring the contacts of persons with serious communicable diseases to identify symptoms sufficiently early to allow prompt isolation and containment of disease.[181] In 1963, Langmuir defined surveillance as the collection, analysis and dissemination of data[182] but by 1968, this definition was extended, as the result of discussions at the 21st World Health Assembly, to include epidemiological surveillance and the responsibility to ensure that effective action was taken, and the application of the concepts to non-communicable diseases. Surveillance is currently defined by the World Health Organization as:

"1. The systematic measurement of health and environmental parameters, recording, and transmission of data.

2. The comparison and interpretation of data in order to detect possible changes in the health and environmental status of populations."[183]

3.2.4 Definition of surveillance

Today, surveillance may have many purposes: to quantify the extent of a health problem, to detect epidemics, to document the distribution and spread of a health problem, to facilitate epidemiological and laboratory research, to test hypotheses, to evaluate control and prevention measures, to monitor changes in infectious agents or in health practice, and to plan for the health needs of a population.[184] It is distinct from *monitoring*[185] which evaluates intervention or action, although the techniques for each may be similar, and from *screening*, which implies that a test of some

form is performed specifically to identify persons at risk. Registers for research purposes are one form of surveillance,[186,187] but have different requirements and purposes compared to a system for the detection of changes in incidence which might accept lower quality case reports in the interests of speed and economy.[185,188] The costs of a surveillance scheme may also be reduced by *sentinel disease surveillance*, which involves only a selected representative sample of eligible respondents.[189]

3.2.5 Requirements of a surveillance scheme

Primary surveillance data collection systems have traditionally been classified as *passive* or *active*. In passive systems, the initiative to report is from the respondent who only notifies positive cases. In active systems, however, the initiative to report is from the surveillance centre: regular reports are requested from respondents, including those who have no positive cases to report. Although both may be affected by incomplete reporting, an estimate of the extent to which the data are non-representative is possible from the latter. The request to report disease by the regular return of a standard postcard was used as early as 1874.[181] The introduction of such active reporting doubled the numbers of reports by primary care physicians.[190,191] Reporting may be affected by distance from a medical facility or the belief of the consumer about what the facility can accomplish[189] and so the value of an individual respondent's contribution should be emphasised. This can be encouraged by feedback which should be regular, relevant and reliable and directed to data providers, decision makers and researchers,[192] by minimising workload[193] and selecting the most

appropriate respondents. For example, moving the responsibility for the reporting of congenital malformations from the mother's to the newborn's physician, and having a centralised single hospital medical records person to review the reports, improved the notification of malformations with a false-positive rate of only 5%.[194] A scheme is not likely to succeed unless it has independence from political or commercial pressures and a totally committed person or group of people at its core.[195] The success of a surveillance scheme is heavily dependent on a clear case definition[196] and a well-defined population under study.[187] Apparent epidemics may reflect changes in registration practices, as for example in the reporting of CDH and limb anomalies.[197] While high sensitivity and specificity in a surveillance scheme are both desirable, one is often at the expense of the other; a specific scheme is more likely to be accurate.[193] Consistency of definitions should be used to allow comparison to other data sources, for example, one difficulty with some international comparisons is the differing definitions of a live birth.[185]

3.2.6 Ethical considerations

Epidemiologists and ethicists have recently collaborated in the formulation of ethical principles for epidemiology[198] and many ethical issues confronting surveillance are similar to those of epidemiology as a whole, in particular the issues of confidentiality and individual privacy. It would be unethical to request participation in a study that is not likely to produce meaningful results or further scientific knowledge for the good of society. Names and other personal identifiers are necessary for two principal related

purposes:[199] for the follow up of individuals and to link data systems. When identifiers are justified, these should be scrambled and the files relating identifiers and individuals maintained securely and separately. Identifying data should be destroyed once it has served its linkage function. The collection of identifying data that will not be used should be avoided. Only aggregated data should be published which precludes identification of individuals but wide dissemination of results is essential. In addition, a scheme should be regularly evaluated to justify its continued existence.[200]

3.2.7 The British Paediatric Association Surveillance Unit

The British Paediatric Association Surveillance Unit (BPASU) was established in 1985 to "improve the surveillance of...conditions in children that could not be monitored through existing data collection systems".[201] The reporting base comprises members of the British Paediatric Association and members of the Faculty of Paediatrics of the Royal College of Physicians in Ireland. Members are asked to report cases of specific conditions (usually 12) seen in the preceding month by returning a monthly reporting card. A card return rate of more than 90% has been reported.[201-204] Although originally established for more rare conditions of childhood, the BPASU reporting scheme has also been used to determine the prevalence and incidence of more common conditions such as diabetes mellitus[205] and in view of this was considered appropriate for estimating the prevalence of treatment for CDH. Since orthopaedic surgeons treat CDH, and both consultant paediatricians and orthopaedic surgeons treat hip instability,[83,206] a parallel scheme for orthopaedic surgeons was established

to allow children treated by orthopaedic surgeons to be reported.

3.2.8 Case definition and study period

In evaluating the CDH screening programme, three groups of children are of interest: those who would develop normal hips without treatment and are treated unnecessarily (false positives); those who would develop a dislocated hip and are not detected by screening (false negatives or failures of detection); and those who would require surgery after the failure of early conservative management (failures of early treatment).

A case definition needed to be clear and precise, both to provide reliable data, and to avoid confusion which might dissuade cooperation. Reports of all initially presumptive positive infants, irrespective of subsequent diagnosis, would have been of interest in measuring the extent of clinical workload but would have imposed too great a reporting burden on clinicians and might have resulted in under reporting. Thus treatment was chosen as a proxy for definitive diagnosis.

Paediatricians and orthopaedic surgeons were requested to report *all infants known to them in whom abduction splinting (including double nappies) for CDH was initiated during the preceding month.* In addition, surgeons were asked to report *infants and young children aged 5 and under receiving a first operative procedure for CDH in that month, with or without general anæsthesia* (Appendices 3.3 and 3.4). From these data, inferences about the false positive rate and an estimate of the false negative rate respectively

could be made. Criteria for exclusion of cases with CDH secondary to another condition were not listed so that the case definitions were as simple and clear as possible. Reports of a first operative procedure in children aged 0-4 years inclusive were requested, since routine births data is supplied in 5-year age bands. However, the protocol card specified children 'aged 5 and under' to ensure children in the fifth year of life were clearly included in the case definition.

A three-month reporting period for cases of abduction splinting and a twelve-month reporting period for cases of a first operative procedure were chosen. Each reporting period was subsequently extended for a month as the BPASU initially reported a lower card return rate for April 1993.[207] Assuming a prevalence of splinting of 10 per 1000 births and a prevalence of a first operative procedure of 1 per 1000, approximately 2000 cases of abduction splinting and 800 cases of a first operative procedure were expected over the reporting periods, corresponding to approximately one case per surgeon or paediatrician.

This study was approved by the Great Ormond Street Hospital and Institute of Child Health ethics committee, contingent on there being no direct contact with the children and their families.

3.2.9 Development and maintenance of the orthopaedic reporting base

Development

In January 1993, a list of consultant orthopaedic surgeons currently practising in the UK and Irish Republic was collated from the British Orthopaedic Association (BOA) and the British Society of Children's Orthopaedic Surgery (BSCOS). Prior notice of the study was provided by an article (Appendix 3.5) in the newsletter of the BOA, the *British Orthopædic News*. Surgeons were sent details of the study, and requested to complete and return a form indicating whether they ever treated children. A reminder letter and second form were sent after three weeks, and the secretaries of non-respondents were telephoned after a further two weeks. Surgeons who treated children were eligible for inclusion in the surveillance scheme and a surgeon was included if there was doubt about his or her eligibility. If the information was not volunteered, secretaries were asked to name any orthopaedic consultant in the department specialising in the treatment of children. New surgeons thus identified were added to the reporting base. All requests for information throughout the study could be sent to a FREEPOST address.

Maintenance

The reporting base was updated throughout the study period from information regarding recent consultant appointments provided by the BOA, and by monitoring advertisements for consultant orthopaedic posts in the *British Medical Journal* and the *Lancet*. For each consultant post advertised, the relevant personnel department was contacted to ascertain

whether the job description included a paediatric workload. Successful candidates for posts with an anticipated paediatric workload were subsequently contacted with details of the study and reporting scheme, and asked whether they were responsible for treating children with CDH. Those eligible were added to the reporting base and asked to report any children treated at any time during the study period. Surgeons were removed from the reporting base if retirement, sickness or death occurred during the study period, or if they subsequently informed the study coordinator that they did not treat children with hip instability or CDH. No surgeon requested to be withdrawn from the scheme.

3.2.10 Surveillance methods

Case notification

At the end of each month, from April 1993 to April 1994 inclusive, surgeons were sent an OS reporting card (Appendix 3.6), and asked to complete and return one half, indicating the number of children under their care receiving a first operative procedure for CDH in the preceding month, or that they had nothing to report. The remaining portion was designed to allow the reporting surgeon to record identifying details of any child(ren) notified that month. A copy of the study protocol was included in the first month's mailing (Appendix 3.4). In addition, both surgeons and paediatricians were asked to notify all children treated with abduction splinting for the months April to July 1993 inclusive, the latter on the BPASU card. Paediatricians were notified of the forthcoming addition of CDH to their monthly reporting card in the regular BPA newsletter

(Appendix 3.7) and sent a protocol card (Appendix 3.3). A one page form requesting further details of the child was sent for each case notified. Special reporting arrangements were made for some surgeons with a high CDH caseload. These included providing forms in advance, and/or arranging for a nominated person such as an orthopaedic nurse or physiotherapist to report cases and complete forms on their behalf. In some centres duplicate reporting was minimised by either an orthopaedic surgeon or paediatrician undertaking to report all cases for that centre.

Encouraging response

Monthly reminders were sent to surgeons not returning OS reporting cards, offering a further opportunity to state whether children with CDH were not treated. The BPASU scheme usually sends reminders to only those paediatricians with three consecutive outstanding cards but, for this study, a single reminder requesting notification of children treated with abduction splinting was sent to paediatricians with one or more outstanding cards for April to July 1993 in March 1994. Surgeons received progress reports twice during the study period and, if appropriate, were prompted for outstanding cards and follow up forms. Surgeons who were members of the BSCOS and/or were named as a surgeon responsible for treating children with neonatal hip instability on the screening practices questionnaires, and who returned few or no reporting cards were identified. These surgeons were sent a reminder letter, modified in the light of this information, emphasising the importance of their cooperation with the surveillance study. Regular feedback to paediatricians was provided

through the BPASU quarterly bulletin. At the end of the study, when surgeons were thanked for their contribution, those who had not returned any cards were reminded that the study had taken place and asked to complete and return a form which asked whether they had treated children with CDH by splinting or surgery during the study period.

'Smart' software (Innovative Software, 1986) was used for the databases and to track the return of cards. Data were transferred electronically between the BPASU office and the study coordinator between April and July 1993, but subsequently the latter coordinated the mailing of the orthopaedic reporting cards and data entry.

3.2.11 Validation of the orthopaedic and paediatric reporting bases

The OS and BPASU reporting bases were compared to lists of consultants compiled independently as part of the orthopaedic and paediatric manpower censuses carried out in September 1992. Details of the surname, initials, hospital, health district and region of each surgeon and paediatrician listed in the manpower census data set were provided as a comma delimited ASCII file. The initial cross-reference was made on the basis of surname. Records with duplicate surnames were checked by hand and matched by initials.

Consultant orthopaedic surgeons and paediatricians identified from the manpower censuses, but not included in the respective surveillance schemes, were sent a form to establish whether they had been a consultant

at any point during the study period (Appendices 3.8 and 3.9), whether this had been at the institution listed in the manpower census, and whether they had treated children for hip instability or CDH during this period. In addition, paediatricians were asked to identify any special interests.

3.2.12 Follow up of notified cases

On notification of a case, a form was sent to the reporting paediatrician or orthopaedic surgeon to obtain background details on treatment and method of detection and to ascertain the eligibility of a case for inclusion in the study. Efforts were made to ensure the forms were as clear and simple as possible, and that the information requested flowed rationally. Each form comprised one side of self-duplicating A4 paper only, to minimise the clinician's workload, and encourage compliance. Self-duplicating paper was used to allow the respondent to keep a copy of the information supplied in case further information was required. The form sent in response to a report of a notified case of abduction splinting was green (Appendix 3.10) and that for a first operative procedure yellow (Appendix 3.11), colours which have been shown to encourage response.[208] Both forms asked for the child's first name and family name, the treating consultant's name, as well several identifiers including the date and hospital of birth and hospital of treatment. This facilitated identification of duplicate reports as well as of the child if additional information was needed. The mother's first name, family name, maiden name and hospital number were requested on the green forms to facilitate subsequent identification of the child because in the early days of life, neonates may

not yet be named or have names that may change. The name and address of the child's general practitioner were requested so that s/he might be contacted where certain information was not available to the reporting hospital consultant. Date of birth was collected to ascertain age at treatment. Details of gender, gestational age, birthweight, ethnic group, side affected, family history, whether first born, mode of presentation, and presence of other abnormalities were collected to characterise children who might be at higher risk of CDH. Ethnic group was not self-reported and since small numbers of non-White children were expected, the OPCS census categories were collapsed for simplicity.

The green splinting form (Appendix 3.10) included 'age suspected' and asked if splinting was preceded by double nappies to explain potential delays between detection and treatment which may result, for example, from a policy of "watchful waiting". Information regarding the means of, and age at, detection of a hip abnormality allowed children treated as the result of a positive screening test to be distinguished from those identified through surveillance, parental concern or especial follow up of children at high risk. The date and type of first operative procedure performed or the date of application of the splinting appliance were needed to confirm eligibility.

3.2.13 Pretesting the follow up forms

Draft copies of the follow up forms were sent for comment to members of the MRC Working Party, the BPASU, and to orthopaedic surgeons who

had expressed an interest in the study after reading the article in the *British Orthopædic News*. A paediatrician and a paediatric orthopaedic surgeon in each health region were chosen to represent a variety of NHS settings, including the community, and teaching and district general hospitals, and clinicians with and without a special interest in CDH. Paediatricians were asked to complete the abduction splinting form only, while surgeons were asked to complete both, in respect of children recently treated. Additional comments on the forms were also solicited. Amendments were made as a result of pretesting and included introducing the term "formal splintage" to distinguish splinting from treatment with double nappies, inserting a direct question on risk factors on the abduction splinting form, and inserting prompts for means of detection on the operative procedure form.

3.2.14 Data entry and coding of forms

All data were entered and verified using a double-entry system (Epi-info v.6, Atlanta) with personal identification data kept separately from the clinical data. An additional summary code for the means of detection was attached (Tables 3.1 and 3.2). The response to each variable was checked for validity and consistency was verified by reference to other variables. In all cases, coding was confirmed by the study supervisor (Dr Dezateux). Differences in coding were exceptional and resolved by discussion. Each form was allocated a case identification number on mailing, using the same system as the BPASU for consistency. This was a combination of the respondent identifier, the type of case and the month of treatment, and an additional number which was allocated sequentially for each month as

cases were notified. For example, the first notification of a first operative procedure was 0027AN/OP/83/01 where 0027AN identifies the consultant: Mr Andrew, OP denotes an operative procedure, 83 the month of April 1993 (the 83rd month of the BPASU scheme) and 01 the first case of the month. The paper forms were filed by case identification number, and thus grouped by respondent for easy reference. Computer files were backed up weekly onto floppy disk and archived on the mainframe computer.

Table 3.1 Coding frame for means of detection of children who received a first operative procedure

10	by routine screening in hospital or on birth examination
20	routine community child health surveillance
30	abnormality suspected by parents or other family member
40	incidental finding when seen in hospital for another condition
50	in consequence of a programme of follow up for children at high risk
99	not known

Table 3.2 Coding frame for means of detection of children who received abduction splinting

	Detected before 3 months age
10	by routine screening
14	incidental finding when seen in hospital for another condition
15	by screening, but delay of more than 2 weeks between detection of abnormality and application of splinting appliance
	Detected after 3 months of age
20	by routine surveillance
26	in consequence of a programme of follow up for children at high risk
30	abnormality suspected by parents or other family member at any age

3.3 VALIDATION OF CASE ASCERTAINMENT

3.3.1 Background

While the high compliance of paediatricians with the BPASU scheme suggested that active reporting might be reliable, complete case ascertainment was not a realistic expectation. Although a high card return rate suggests good compliance it carries no guarantee of completeness of ascertainment or the accuracy of returns. Furthermore, in recent years there has been increased recognition of the need for multiple sources of ascertainment to increase the accuracy of prevalence estimates. Means of validating the active reporting by paediatricians and orthopaedic surgeons were sought.

3.3.2 Capture recapture analysis

Capture recapture analysis derives its name from its development and use to estimate the size of wildlife populations, whereby animals are captured, marked,

released and subject to recapture. However, it is increasingly used in epidemiological studies,[205,209-214] and may also be referred to as "ascertainment intersection"[215] or a "comparison of multiple methods of ascertainment".[216] The technique relies on case ascertainment from two or more independent sources and uses the proportion of cases found in common with other sources to generate estimates of missing cases and the total affected. Several assumptions should hold for the method to be valid:[213]

1) there is no change to the population during the investigation (the population is closed)

2) identification of individuals is such that all and only true matches can be made

3) the samples are independent

4) there should be no 'variable catchability', ie. within source variation in probability of ascertainment of cases.

3.3.3 Validation of the cases of abduction splinting

Members of a group which had been convened to discuss modifications to the OPCS National Congenital Anomaly System (formerly the OPCS Monitoring Scheme for Congenital Malformations)[174] were asked by postal questionnaire (Appendix 3.12) whether they were aware of any sources of data of children treated for CDH, including regional congenital malformations registers, and if so, for the name of the source and a contact name and telephone number.

3.3.4 Validation of the cases of a first operative procedure

3.3.4.1 Validation areas

Hospital episode system (HES) data were judged a suitable source of

validation for the operative procedure cases, but as discussed earlier (section 3.2.2), this could not be undertaken nationally since verification of HES-identified cases by reference to the medical records was essential.[180] Geographical areas for validation were selected. The former Northern and South Western regions were chosen because they were geographically diverse and because a researcher in each region had previously undertaken work of this kind. These areas account for 10.2% of UK births (1993 figures). When the researcher in the South Western region became subsequently unavailable, Wessex was substituted but an additional area was needed to maintain a similar population coverage. Scotland was chosen because it was feasible for the researcher from the Northern region to visit the Scottish centres. Furthermore, episode data from Scotland were linked which would facilitate identification of *first* operative procedures. The final choice of validation areas accounted for 18% of UK births (1993 figures).

3.3.4.2 Identification of cases of a first operative procedure from HES data

HES data are collected in order to provide basic details for every episode of every patient's stay in hospital, where an episode is considered the period from admission to and discharge from a single specialty. It was essential that cases were identified from HES data using selection criteria equivalent to the definition used for the OS reporting scheme (chapter 2). The recorded variables of interest in this study were date of birth, hospital of treatment, case records number (CRN), period of episode, main diagnoses, and the date and type of the first operative procedure undertaken.

3.3.4.3 Case definition

In the Wessex and Northern regions, a list of episodes occurring to children aged 5 and under during the period 1/4/93 to 30/4/94 with the appropriate International Classification of Diseases, Injuries and Causes of Death (ICD) version 9[217] or OPCS operative procedure codes[218] was requested from the regional statistician. The ICD version 9 code for CDH was 754.3x and the OPCS operative procedure codes were X22x, X48x and X49x (a correction of congenital deformity of the hip, immobilisation using a plaster cast, or other immobilisation, respectively). The study office made the request for the Wessex data and these were provided on a floppy disk in ASCII format, but the Northern data were sent in paper form directly to Dr Hey who made the initial request to the regional statistician. In Scotland these data were provided on disk by the Information and Statistics Department (ISD) of the Common Services Agency of the Scottish Health Service. The Wessex file supplied each child's date of birth, date of admission, start date of episode, end date of episode, up to 6 diagnoses, 4 operation codes, date of first operation, the district health authority of the child's residence, the provider code and the main speciality code. The Northern file supplied the above, apart from the date of operation and district of residence and the date of discharge was given, rather than the start and end of episode dates. In addition, the Northern file included the case record number, consultant identifier and child's postcode. The Scottish file supplied similar data to that provided by the Northern region, apart from the child's postcode, but additionally provided all episodes for that child which had occurred within the Scottish Health Service.

3.3.4.4 Preparation of HES data

The medical records manager of each hospital listed for Wessex was approached to ascertain the children's case record numbers in order that the medical records might be located. The Wessex data and Scottish data were examined to identify children who were ineligible, for example, because CDH was secondary to another condition. Children were excluded from the Scottish list if episodes of treatment for CDH were listed which had occurred before the start of the study period.

3.3.4.5 Ethical approval

The study was approved by the Great Ormond Street Hospital and Institute of Child Health ethics committee (section 3.2.8). Permission for the identification of records though HES data was supplied by the Northern Regional Health Authority, the Wessex Regional Health Authority and the Information and Statistics Department of the Common Services Agency of the Scottish Health Service. In the Northern region, each health district or trust granted access to the medical records for this study to Dr Edmund Hey, a member of the MRC Working Party, as part of the Northern Regional Fetal Abnormality Survey. On 3 occasions, written confirmation of ethical approval and details of the current study were requested by medical records officers in the Northern region. Medical records officers in each Scottish hospital were supplied with written details of the study and of the ethical approval obtained. No further information was requested from hospitals in Scotland. In the Wessex region, details of the study were provided by post and telephone to the medical records officer for each hospital. In three hospitals, permission to access the medical records was sought and obtained by the medical records officer from the relevant clinicians, in two chairman's action was taken, and in the sixth, submission of an ethics form was requested and supplied.

3.3.4.6 Design of validation forms

Information was required for each child to determine whether s/he had been eligible and whether s/he had been reported previously to the OS scheme. Sufficient additional information was requested in order to characterise children who were not identified though the OS scheme. A copy of the form is attached (Appendix 3.13). While similar to the yellow forms described earlier, modifications were made to take account of the constraints of completing a form from written records alone. The child's previous surname was requested in case of name changes. All operative procedures before the end of the study period were requested so that eligibility could be determined by the study coordinator, rather than by the peripatetic researchers. Details of periods of traction were requested to help to explain time differences between detection of abnormality and first operative procedure. The results of the last X-ray or ultrasound examination before surgery were requested. The questions relating to the age at detection were tailored to allow the researchers to transpose separate items of information directly from the medical records to the forms, rather than to take a decision regarding the overall picture. The results of the neonatal and six-week examinations were requested explicitly; this information had sometimes been provided additionally on the yellow forms.

3.3.4.7 Training

A training day was organised for the two validation researchers, one a research nurse, the other a physiotherapist, both of whom were familiar with medical records, medical terms and CDH. Two other members of the MRC Working

Party also attended. This training day was to encourage commitment to conscientious completion of the forms and to discuss the content of and possible amendments to the forms. The study aims and design were described and two forms were completed by each person from the medical records of children treated for CDH at Great Ormond Street Hospital (GOSH), but who were not eligible for inclusion in the study. These were provided by Mr Fixsen, a consultant orthopaedic surgeon at GOSH. At least two people independently completed a form for each child to test the reliability of the form. The forms were found to be reliable.

3.3.4.8 Data storage and entry

Each researcher photocopied completed validation forms before sending them to the study coordinator in case they went astray in the mail. Forms were returned in batches as each hospital was visited. All data were entered with a double-entry system (Epi-info v.6, Atlanta) with personal identification data kept in separate files from the clinical data.

The forms were categorised and the complexity of first operative procedure was classified as per the OS scheme categories. Yellow OS scheme forms and validation forms were matched by child's name and date of birth. Details of all the eligible children identified by the validation exercise and similar details for children reported to the OS scheme not identified by HES were contained in one file, with an additional variable to indicate the 3 groups (table 3.3).

Table 3.3 Classification of children grouped according to means of case ascertainment

Group	Description
1	reported to OS scheme and identified in HES
2	identified in HES only
3	reported to OS only

Calculation of the ascertainment-adjusted total number of cases (N) and appropriate confidence intervals was as follows, using the adjusted maximum likelihood estimator for small samples.[219]

(Equation 3.1)

$$(N) = [(OS+1)(HES+1)/(both+1)]-1$$

where

OS = number of cases reported to OS scheme

HES = number of cases identified in HES data

both = number of cases reported to OS scheme and identified in HES data

(Equation 3.2)

$$Var\ (N) = ((OS+1)(HES+1)(OS-both)(HES-both))/((both+1)^2\ (both+2))$$

and 95% confidence intervals were constructed by $N \pm 1.96\ (\sqrt{Var(N)})$.

4: RESULTS

4.1 SURVEY OF SCREENING AND MANAGEMENT

4.1.1 Response

Questionnaires were sent in the first instance to 267 named paediatricians. In 20 units, a questionnaire was sent to a second paediatrician, if a more appropriate respondent was suggested, or if the first questionnaire was not returned. Twenty-two additional units were identified by respondents and 12 units had closed, amalgamated, or did not provide neonatal care.

Information was provided for 254 (92%) of the maternity units identified. Responding units (737,906 total births per annum) tended to be responsible for more births than non-responding units (56,165 total annual births); within each region, the mean difference (95% CI) was 610 births (115, 1105). The number of births in the UK and Republic of Ireland in 1992 was approximately 832460,[220] of which 98% were delivered in NHS hospitals.[221] This suggests that the maternity units identified by this survey covered 95% of annual births, on which information was obtained for 93%. Additional information on the use of ultrasound was requested from radiologists in 79 units (99% response) and on the management of infants with dislocated or dislocatable hips from orthopaedic surgeons in 144 units (86% response).

As the questionnaire was designed to allow a 'don't know' or 'not applicable' response where necessary, the denominators in the results section excluded units

where no information was available or a question was not applicable and the number of responding units is given. The results were expressed as percentages with units, rather than births, as the denominator. This would distort the overall conclusions only if small units were likely to be consistently different to large units. The replies to the two questions which asked if ultrasound was available and whether a high risk policy was in place were examined using births as the denominator. The mean number of births reported in units using ultrasound was 2962 (95% CI: 2753, 3171) which was comparable to the mean number of births in centres not using ultrasound (2865 (95% CI: 2543, 3187)). The mean number of births reported in units with a high risk policy was 3003 (95% CI: 2766, 3240) which was comparable to the mean number of births in units without a high risk policy (2782 (95% CI: 2529, 3035)).

4.1.2 Screening by clinical examination

The first screening examination was routinely offered within 24 hours of birth as recommended in the current guidelines, in only 69% of units, but in all units within 48 hours. In 30% of units, a second routine screening test was never undertaken before discharge; age at discharge was the determining factor, and only 21 units (8%) attempted a second examination in all infants. A paediatric senior house officer was responsible for routine screening before discharge, either alone (182 units; 72%), or together with other junior paediatric and obstetric staff (62 units; 24%). Formal training with a 'Baby Hippy' hip simulator model was available in 95 units (37%). Clinical or beside teaching formed the basis of most training, supplemented with a video or lecture in 17 (7%) units.

4.1.3 Use of ultrasound

At the beginning of 1994, 176 units (69%) had access to ultrasound examination of the hips compared to 14% in 1984 (Figure 4.1). A further 4% planned to introduce this service over the next 12 months. Ultrasound was usually performed to assess or manage infants with clinically detected hip abnormalities, or to screen infants at high risk of CDH, and only three units undertook routine universal primary ultrasound screening (table 4.1A). Radiologists and radiographers, either alone or with orthopaedic surgeons, performed and reported the ultrasound examination (table 4.1B). The type of examination performed and method of reporting varied between units. Some units performed a static examination, some dynamic examination, and some a combination of both (table 4.1C). Of those units performing a static examination, most (61%) used the Graf technique or similar technique involving the measurement of angles, in their reporting. The person responsible for the hip ultrasound service was usually a radiologist, (96 units; 87%) or an orthopaedic surgeon (10 units; 12%).

Figure 4.1 Percentage of maternity units in the UK and Irish Republic with access to ultrasound imaging, by year of introduction

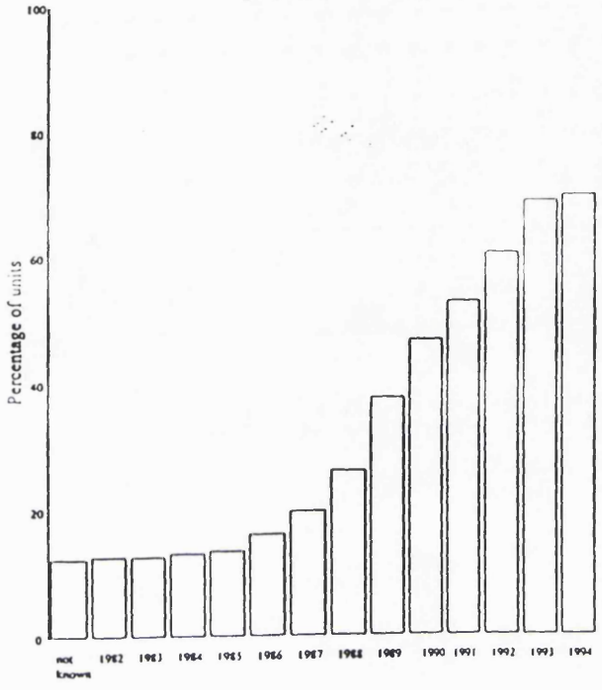


Table 4.1 Use of ultrasound (176 units)

A. Children referred for ultrasound assessment	number of units (%)
clinically detected hip instability alone	79 (45)
high risk alone	9 (5)
both clinically detected hip instability and high risk	85 (48)
all children	3 (2)
B. Speciality of examiner performing ultrasound	
radiologist/radiographer alone	125 (71)
radiologist/radiographer and orthopaedic surgeon	28 (16)
other	23 (13)
C. Type of ultrasound performed	
static alone	39 (22)
dynamic alone	60 (34)
both static and dynamic	67 (38)

4.1.4 High risk infants

These questions aimed to ascertain the extent to which units had a different policy for the screening and management of high risk infants, irrespective of the clinical findings at the O-B test. However, while a high risk policy was reported in 159 units (63%), in 40 of these the question regarding whether this applied to infants who were clinically normal was answered 'no' or 'don't know' or left unanswered. Thus less than one half of units (119; 47%) clearly operated a high risk policy. The criteria used to identify high risk infants varied between units (Table 4.2). Infants with some or all of these risk factors were further assessed with ultrasound in all units where it was available (85 units; 53%) and/or a repeat clinical examination (125 units; 79%).

Table 4.2 Criteria used to identify infants at high risk of CDH (n=119 units)

risk factor	% units
family history of CDH	100
breech presentation	84
talipes	66
other postural abnormalities	42
oligohydramnios	31
family history of clicky hips	16
caesarean section	9
intrauterine growth retardation	5

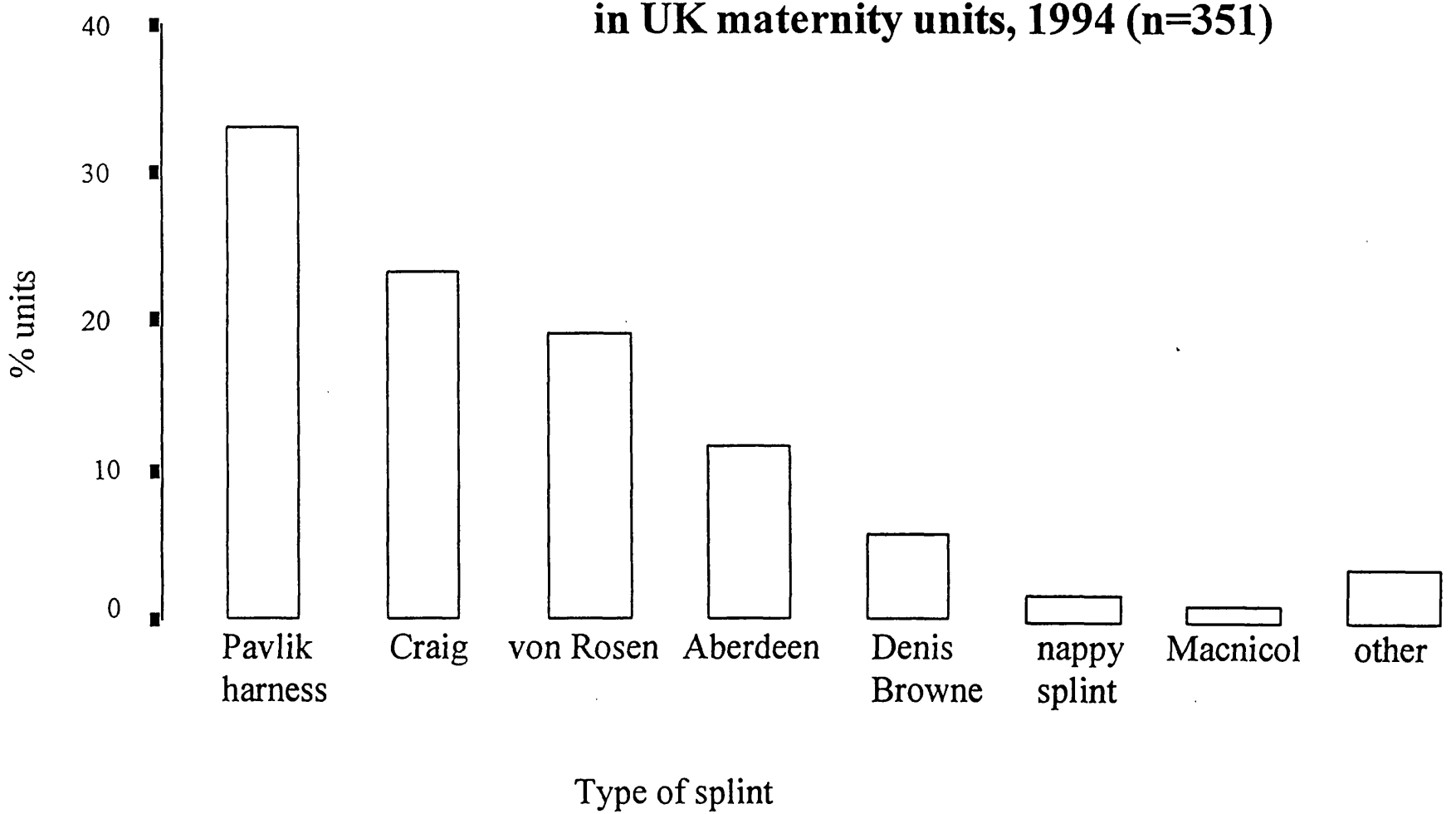
4.1.5 Management

Infants in whom a dislocated or dislocatable hip was suspected before discharge were reported to receive immediate treatment in one third and one

quarter of units, respectively (questions 15a and 15b). However, a plastic splint was fitted before further clinical or ultrasound examination in only 2 units, which suggests that strictly immediate treatment was fairly unusual. Orthopaedic surgeons were primarily responsible for making the decision to start treatment (237 units; 94%), but the timing, type and duration of treatment provided varied.

While initial treatment for dislocated hips was usually a splint appliance (225 units; 94%), in 13 units (6%) this was a plaster of Paris cast. Dislocatable hips were treated initially with a splint appliance in 214 units (89%), with a plaster of Paris cast being used in only 3 units. A total of 15 different types of splint were used as initial treatment (Figure 4.2), with a Pavlik harness the most frequently used. Reported duration of treatment typically ranged from 2 to 52 weeks, but was 12 weeks or less in 70% of units. Splint appliances were fitted by orthopaedic surgeons, or by physiotherapists, orthopaedic nurses, orthotists, plaster room technicians, or appliance officers usually under the supervision of a consultant orthopaedic surgeon. Ultrasound was used to monitor the progress of a child's hips in abduction splinting in 116 units (66%), and to determine treatment duration in 62 (38%) of the 176 units with access to ultrasound services.

Figure 4.2 Frequency of splint types used in UK maternity units, 1994 (n=351)



Further assessment was arranged for infants with a dislocated or dislocatable hip that stabilised spontaneously without treatment in 237 units (90%). This included a pelvic X-ray, usually at between 3 and 6 months of age, in 111 units (52%), as well as clinical follow up which varied in duration from 3 months to weight-bearing age and beyond. Double nappies were used in 89 units (36%): to treat established dislocation in 6 units, to treat unstable hips in 33 units, to treat 'clicky' hips (not defined in the questionnaire) in 20 units, but primarily as an interim treatment until diagnosis was confirmed (67 units; 75%). In addition, they were used for 'weaning' (after a period of abduction splinting in a plastic splint), and for preterm and other infants where a plastic splint was considered too large.

A consultant orthopaedic surgeon was named as the person primarily responsible for the management of infants with dislocated or dislocatable hips in 222 units (94%). Only 40% of respondents identified an individual in their district, usually a consultant paediatrician (32%) or orthopaedic surgeon (36%), responsible for keeping the screening programme under review.

4.2 SURVEILLANCE STUDY

4.2.1 The orthopaedic reporting base

The initial questionnaire to identify surgeons with a paediatric workload was returned by 749 (69%) of surgeons within three weeks of the first mailing. A total of 82% had replied two weeks after the postal reminder, leaving 194 non-respondents whose secretaries were telephoned. Of the 1225 surgeons identified, 517 (42%) indicated that they treated children and were included in the Orthopaedic Surveillance (OS) scheme. The number of orthopaedic consultants

identified through the various sources and the number who treat CDH are given in Table 4.3. The orthopaedic manpower census identified 1047 current consultant posts, of which 4 were vacant. Of the 60 surgeons named in the manpower census but not included in the OS scheme, 7 did not reply to the postal questionnaire, 32 were consultant orthopaedic surgeons but had not treated children with CDH, and 4 were not consultant orthopaedic surgeons in clinical practice during the study period. This suggested that only 3% (n=17) of surgeons responsible for treating children for CDH were not included in the OS scheme.

Table 4.3 Source from which surgeons were identified for inclusion in the orthopaedic surveillance scheme

Source	Number	Number who treat children with CDH
Home Fellow[a] of the BOA[b] in January 1993 with a current address in the UK or Irish Republic	1086	453
nominations from other surgeons, and successors of retiring surgeons	9	7
new Home Fellows	79	25
surgeons identified by secretaries during telephoning	7	7
opportunistically	4	4
new appointments advertised in *British Medical Journal* and *Lancet*	40	21
TOTAL (%)	1225 (100)	517 (42)

[a]Home Fellow: consultant orthopaedic surgeon practising in the UK or Irish Republic
[b]British Orthopaedic Association

4.2.2 The paediatric reporting base

The paediatric manpower census identified 1130 current consultant posts, of which 909 were hospital-based, 221 community-based, 1 both, and 61 either vacant or occupied by a locum. Of the 129 paediatricians listed in the manpower census but not included among the 1179 paediatricians in the BPASU scheme during the study period, 17 did not return the postal questionnaire and 14 were not in clinical practice in the UK during that time. This suggested that 8% of eligible paediatricians were not included in the BPASU scheme. Three of six paediatricians who reported starting treatment for hip instability during the study period specified that such children had been referred to an orthopaedic surgeon.

All three of these surgeons were included in the OS scheme. Of the paediatricians not included in the BPASU, 41% reported a special interest in community child health or neurodevelopmental assessment, and 24% in neonatology (Table 4.4).

Table 4.4: Reported special interest(s) of paediatricians not included in the BPASU scheme during the study period

Special interest(s)	Number of paediatricians (%)
Neonatology	23 (24)
Community child health or neurodevelopment[a]	40 (41)
Other[b]	27 (28)
None/not specified	8 (8)

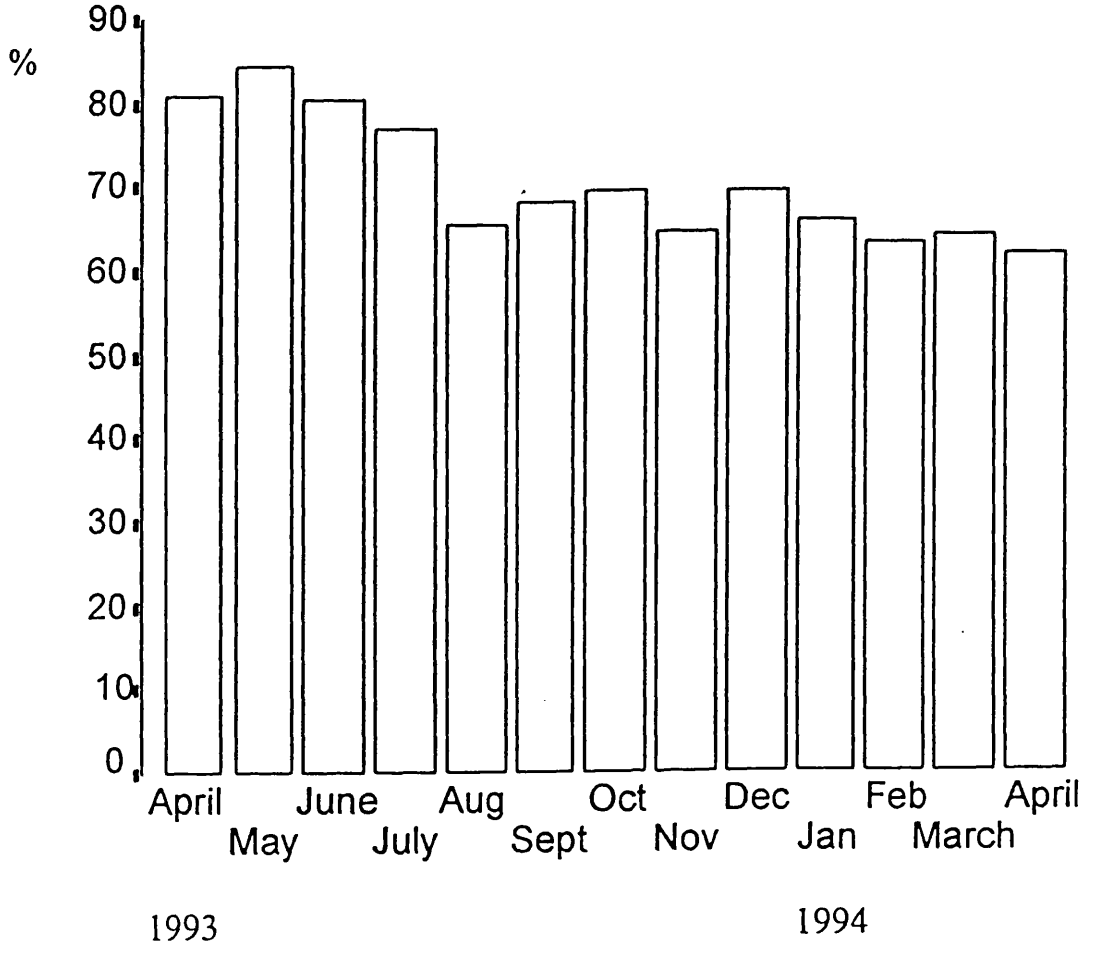
[a]includes special needs assessment, child abuse

[b]includes sub-specialties such as respiratory medicine, oncology, endocrinology, immunology, infectious disease, gastroenterology, intensive care

4.2.3 Compliance with the reporting schemes

Each month, approximately 50% of OS reporting cards were returned within 3 weeks of mailing and the mean (median) monthly card return rate over the 13 month period was 72% (92%). A significant decrease in card return was seen over time (Figure 4.3) (test for trend in proportions,[222] $p<0.01$). No cards were returned by 37 surgeons, of whom one later confirmed he had treated children with abduction splinting and with surgery, one had treated children only with abduction splinting and one had treated children only with surgery during the study period.

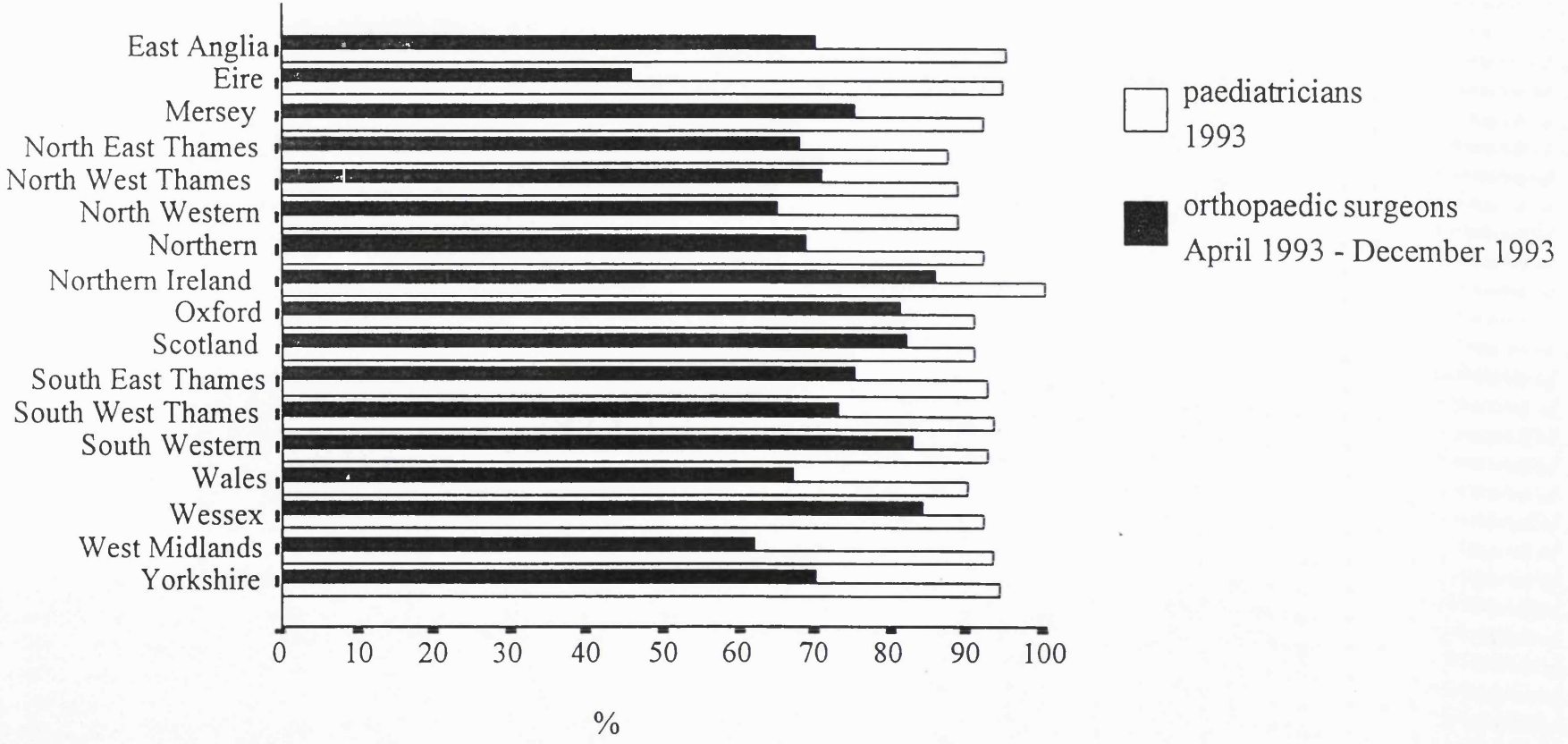
Figure 4.3 Monthly card return rate of Orthopaedic Surveillance scheme



Direct comparison of card return rates between the OS and BPASU schemes was possible for the period April to June 1993 only, when the published card return rate for paediatricians was 91%[207] and the corresponding figure for surgeons was 83%. The reminder to paediatricians for cases of CDH increased the percentage of returned cards to 96%. When examined by country or former NHS region, the OS scheme card return rate showed marked variation (Figure 4.4), ranging from 70% for the Northern to 91% for the Mersey region. The reported variation in the BPASU card return rate[207] was less marked, ranging from 88% for North East Thames to 100% for Northern Ireland. There was no apparent concordance in the geographical variation in card return rates between the two schemes (Wilcoxon matched pairs signed ranks test $p<0.001$).

Owing to the low card return rate, cases reported by paediatricians and orthopaedic surgeons from the Irish Republic will be considered separately.

Figure 4.4 Percentage of reporting cards returned, by country or former NHS region



4.2.4 Cases of abduction splinting

Prevalence

A total of 944 forms were sent in response to notifications of children treated by abduction splinting, 346 to paediatricians and 598 to surgeons, of which 684 (72%) were returned. Of the questionnaires returned, 150 cases were not eligible for inclusion in the study and 71 were duplicates of previous reports. Most (70%) duplicate reports were the result of a paediatrician and a surgeon reporting the same child. Cases were ineligible for several reasons (Table 4.5) but primarily because the splinting appliance was first applied outside the study period. Children with hip dislocation secondary to another disorder, such as cerebral palsy, or with hip dislocation as part of a syndrome, for example, arthrogryposis were excluded.

Table 4.5 Reasons for ineligibility for notified cases of abduction splinting

Reason for ineligibility	Number of cases
Child splinted outside study period	58
Notification error	45
Child received surgery, not splinting	37
Instability resolved without treatment	5
Child could not be identified	4
Acquired hip dislocation	1
TOTAL	150

Forms were not returned for 34 (37%) of notified cases of abduction splinting in the Republic of Ireland. The 57 children who were born and treated in the

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Forms were not returned for 34 (37%) of notified cases of abduction splinting in the Republic of Ireland. The 57 children who were born and treated in the

Republic of Ireland will be discussed separately.

From these confirmed cases, the prevalence of abduction splinting in the UK was 406/258 300 per 1000 live births = 1.6 (95% CI: 1.4, 1.7). If all notified but unconfirmed cases were true cases, the prevalence of abduction splinting would have been 2.4 (95% CI: 2.3, 2.6). However, 32% of returned forms were ineligible or duplicate reports. If it is assumed that a similar proportion of the unreturned forms were not true cases, the prevalence of abduction splinting would be 1.9 (95% CI: 1.7, 2.1).

Characteristics of confirmed cases of abduction splinting

There was an excess of girls (Table 4.6) compared to the female:male ratio of live births in England and Wales (E&W) in 1993 of 1:1.06.[223] There was a statistically significant excess of children with a birthweight over 3.5kg compared to the babies born in E&W in 1993[221] (47% and 39% respectively; test for difference between proportions, $p<0.001$).

In E&W in 1993, 68% of births were to married women,[223] of which 39% of these births were first born, which is comparable to the percentage of first born children in this study. Most (95%, 95% CI: 93%, 98%) children were White. The proportion of infants born in 1993 who were White, is not available (Dave Canham, Office of National Statistics, personal communication) but in 1993-5, 90% of children aged 0-4 in Great Britain were White,[220] and assuming that the proportion of White births is unchanged, this suggests that the prevalence of abduction splinting was significantly higher in White children.

Table 4.6 Characteristics of confirmed cases of abduction splinting

Characteristic	
female:male ratio	4:1
median (IQR) birthweight	3.46 kg (3.10 to 3.79)
median (IQR) gestational age	40 weeks (39 to 41)
first born	145 (38%)
ethnic group	
White	313 (95%)
Asian	9 (3%)
Oriental	3 (1%)
other	3 (1%)

Data items were complete for all 406 confirmed cases, with the exception of birthweight (85% complete), gestation (86%), ethnic group (81%) and risk factors (94%). Missing data were actively sought from reporting clinicians and, and with their consent, from general practitioners.

At least one characteristic thought to predispose a child to CDH was reported in 184 (48%) of children. The distribution of these risk factors is shown in Table 4.7.

Table 4.7 Reported risk factors for confirmed cases of abduction splinting

Risk factor	Number[a] (%) of cases
positive family history of CDH	66 (17)
breech presentation	88 (23)
positional deformity of lower limb	24 (6)
oligohydramnios/multiple birth	2 (0.5)
torticollis/scoliosis/sacral dimple	5 (1)
growth retardation	2 (0.5)
caesarean section	5 (1)
ligamentous laxity	1 (0.3)

[a] n=381

In nearly half (48%) of children, the left hip alone was affected, but in more than one third (37%) both hips were affected.

Geographical distribution of cases of abduction splinting

The prevalence of reported cases of abduction splinting ranged from 0.26 per 1000 live births in Northern Ireland to 3.99 in Wessex (Table 4.8). This geographical variation was significantly greater than might be expected by chance ($\chi^2 = 230$, $p<0.001$) and may reflect variable case ascertainment, as well as variation in treatment. There appeared to be little association between the regional prevalence of confirmed cases of abduction splinting and the regional card return rate (Pearson's correlation coefficient, $\rho = 0.14$) suggesting that the variation in prevalence regionally was not explained by variation in the card return rate. Only 4 children received treatment in a region different from their region of birth.

Table 4.8 Geographical variation of confirmed cases of abduction splinting

Region or country birth	No. of cases	Approximate[a] number of births in 4 months[221]	Prevalence of abduction splinting per 1000 live births
Wessex	52	13042	3.99
Wales	37	39626	3.03
Trent	48	20270	2.37
Scotland	44	21100[220]	2.09
South Western	25	13128	1.90
East Anglia	13	8529	1.57
West Midlands	34	23282	1.46
North Western	24	18124	1.32
Oxford	14	11602	1.21
Northern	14	12866	1.09
Mersey	10	10180	0.98
North West Thames	15	16680	0.90
Yorkshire	14	16092	0.87
South West Thames	10	12755	0.78
South East Thames	9	16827	0.53
North East Thames	7	18784	0.37
Northern Ireland	22	83300[220]	0.26
not known	14	-	-
TOTAL	406		

[a] estimated from annual figures

Mode of presentation of cases of abduction splinting

Most (96%) children were detected by the neonatal screening programme, 82% within 48 hours of birth. Ultrasound was used to assess 151 infants (39%), of

whom 83 (61%) were at high risk. Three children were found to have abnormal hips before 3 months of age when in hospital under investigation for other problems and a further 3 were brought to the attention of the health service by their parents. After the age of 3 months, 13 (3.2%) children were detected by routine surveillance and 2 (0.5%) were detected through the follow up of high risk infants.

Diagnostic tests prior to abduction splinting

The decision to treat was made on the basis of clinical findings alone in 200 (53%) children, with the addition of ultrasound imaging in 125 (31%), on the findings of clinical, ultrasound and X-ray examination in 13 (3.4%) and on the basis of clinical and X-ray examination alone in 41 (11%). Ultrasound imaging was undertaken at a median age (IQR) of 8 days (2 to 20) while X-rays were undertaken at a median age (IQR) of 103 days (43 to 170). Twenty children were treated on the basis of an abnormal ultrasound finding despite normal findings on clinical examination.

Treatment of NHI

Double nappies were the initial treatment for 88 (23%) children, and at the time of reporting, 51 of these had not received treatment with a formal splinting appliance. A total of 15 splint types were used but the Pavlik harness was used most frequently in 33% of children treated with a formal splinting appliance.

4.2.5 Cases of a first operative procedure

Prevalence

A total of 556 questionnaires were sent in response to notifications of a first operative procedure for CDH, of which 436 (78%) were completed. Of those completed, 103 cases were not eligible for inclusion in the study, 10 were duplicate reports by the same surgeon, and 4 had been reported already by another surgeon. Cases were not eligible primarily because the date of the first operative procedure fell outside the study period or because a case was reported in error (Table 4.9). A questionnaire was returned for 310 eligible cases, and a further 9 were completed on behalf of a surgeon by the study coordinator (SG). Of these 319 cases, one child was born and treated in the Republic of Ireland and will be considered separately. Forms were not returned for 16 notified cases of a first operative procedure in the Republic of Ireland.

Table 4.9 Reasons for ineligibility of reported operative cases

Reason	Number of cases
first operative procedure not in study period	44
born outside UK and Irish Republic	3
no surgery	10
form not returned because "reported in error"	28
child's details not available because referred	2
child more than 5 years old	5
hips not dislocated	5
CDH as part of a syndrome/neuromuscular disorder/other major malformations	6
TOTAL	103

The number of cases of CDH in a population is usually estimated from observation of a birth cohort and expressed per 1000 live births. In this cross-sectional study, however, the denominator was the children at risk of a first operative procedure for CDH who were aged 5 and under during the 13 month period. These data are not available, but the number of births over a 13 month period was an adequate approximation. Using figures from 1993, this was approximately 825175 births.[220] The prevalence of a first operative procedure based on reports to the OS scheme was calculated:

prevalence of first operative procedure = 318/825175

= 0.39 (95% CI: 0.34,0.43) per 1000 live births.

Characteristics of confirmed cases of a first operative procedure

Data items were complete for all 318 confirmed cases, with the exception of birthweight and gestation (84% complete), ethnic group (89%) and risk factors (90%). Missing data were actively sought from reporting clinicians and, and with their consent, from general practitioners. There was a significant excess of girls (table 4.10), compared to the female:male ratio of all children aged 0-4 years in the UK in 1993 of 1:1.05.[224] There was no statistically significant excess of low birthweight (less than 2.5kg) children (6% and 6%, test for difference between proportions, p=0.26), or children with a birthweight over 3.5kg (42% and 39%, test for difference between proportions, p=0.18) compared to the population at large.[221] A similar proportion of children were firstborn (39%) compared to the proportion (39%) of children born in E&W in 1993to nulliparous married women.[220]

Table 4.10 Characteristics of confirmed cases of a first operative procedure

Characteristic	
female:male ratio	8.4:1
median (IQR) birthweight	3.4kg (3.1 to 3.8)
median (IQR) gestational age	40 weeks (39 to 40)
first born	111 (39%)
ethnic group	
White	266 (94%)
Asian	10 (4%)
Oriental	3 (1%)
other	3 (1%)

In Great Britain in 1993-5, 90% of children aged 0-4 years were White.[220] This suggests that the proportion of White children receiving a first operative procedure for CDH in this study (94% (95% CI: 92%, 97%) was significantly higher than expected.

At least one recognised risk factor for CDH was reported in 94 (33%) of the children for whom data on risk factors were available. The distribution of risk factors is shown in Table 4.11. Among the children not detected by screening, 62 (30%) had at least one risk factor for CDH. While the prevalence of a positive family history was similar in those detected by screening and not detected by screening (12% and 11% respectively), the prevalence of breech presentation was significantly higher among those detected by screening (24%) compared to those not detected (11%).

Table 4.11 Prevalence of known risk factors among cases of a first operative procedure reported to the Orthopaedic Surveillance scheme

Risk factor	Total number (%) of cases[a]	Number (%) of cases not detected by screening[b]
positive family history	33 (11)	23 (11)
breech presentation	44 (15)	24 (11)
positional deformity of foot	10 (3)	4 (2)
oligohydramnios/multiple birth	4 (1)	2 (1)
torticollis/scoliosis/hemivertebrae	5 (2)	3 (1)
caesarean section	7 (2)	4 (2)
plagiocephaly	5 (2)	4 (2)

[a]n=287 [b]n =203

Geographical distribution of cases

The number of confirmed cases per 1000 live births by NHS health region (1993 definition) or country of birth, is shown in Table 4.12 and varied from 0.2

to 1.08 per 1000 live births. This variation is significantly greater than might be expected by chance ($\chi^2 = 75.4$, $p < 0.001$) but is likely to reflect variable completeness of case reporting and referral patterns as well as true differences in prevalence. However, a substantially higher prevalence in Scotland and Northern Ireland was reported, which is consistent with the published literature (section 2.3.1). Of the 294 children for whom hospital of birth was known, 33 (11%) were treated in a different region or country from their region or country of birth, which has implications when assessing the effectiveness of local screening programmes. In particular, cases born in Wales were likely to be treated in an English health region.

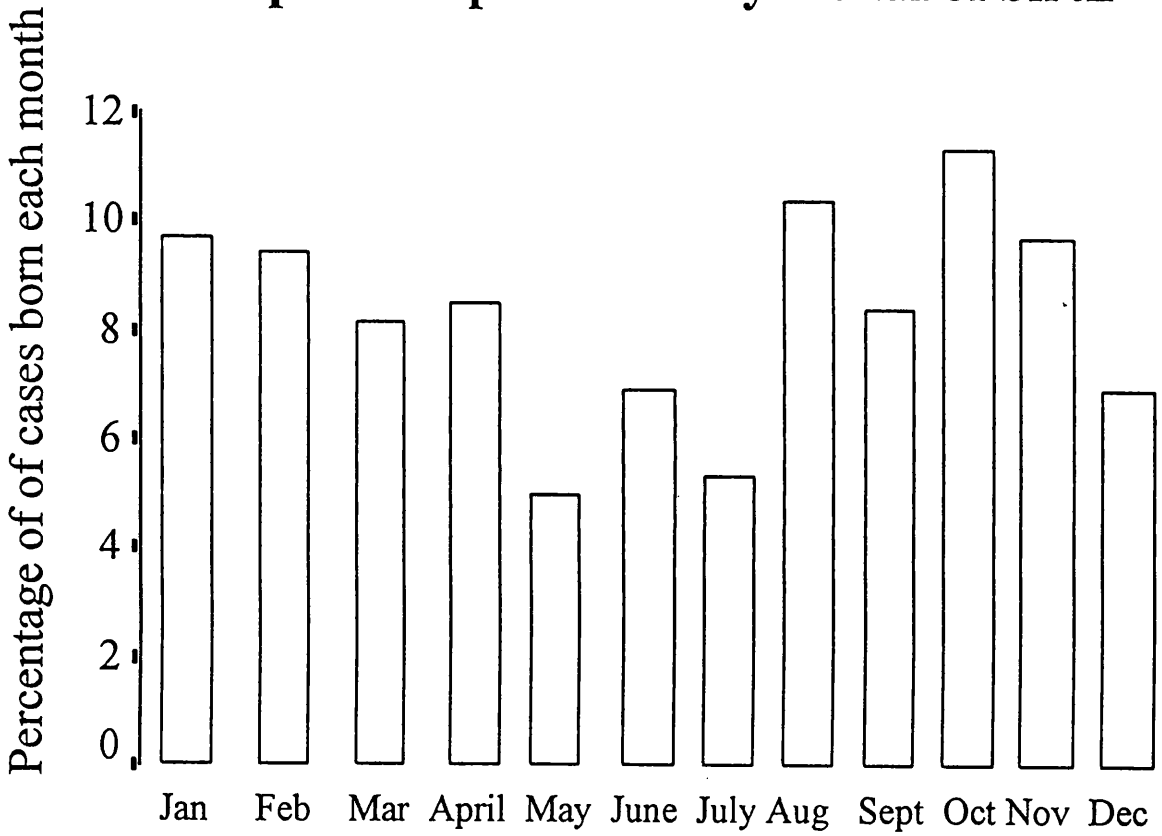
Table 4.12 Geographical distribution of cases

Region or country of birth	Number of cases born	% cases treated in region of birth	Approximate births in 13 months[221]	Cases per 1000 live births
Northern Ireland	29	100	26975[220]	1.08
Scotland	44	98	68575[220]	0.64
Trent	29	97	65876	0.44
North West Thames	22	86	54209	0.41
Wessex	17	94	42385	0.40
Northern	16	88	41814	0.38
East Anglia	10	100	27718	0.36
Wales	15	29	39626	0.35
Mersey	11	91	33084	0.33
South West Thames	13	92	41453	0.31
Yorkshire	15	80	52298	0.29
South Western	12	75	42666	0.28
West Midlands	17	88	75666	0.22
Oxford	8	100	37705	0.21
North Western	13	100	58902	0.21
South East Thames	11	82	54688	0.20
North East Thames	12	83	61047	0.20
not known	24	-	-	-
TOTAL	318	89		

Seasonality

It has been suggested that children born in the winter months are at higher risk of CDH[225-228] but no clear trend was seen in the distribution of cases by month of birth (Figure 4.5) and a test for a seasonal trend using a Kolmogorov-Smirnov type statistic[229] was not statistically significant (p=0.62).

Figure 4.5 Distribution of reported cases of a first operative procedure by month of birth



Hip affected

In nearly two-thirds of children, the left hip alone was affected (Figure 4.6).

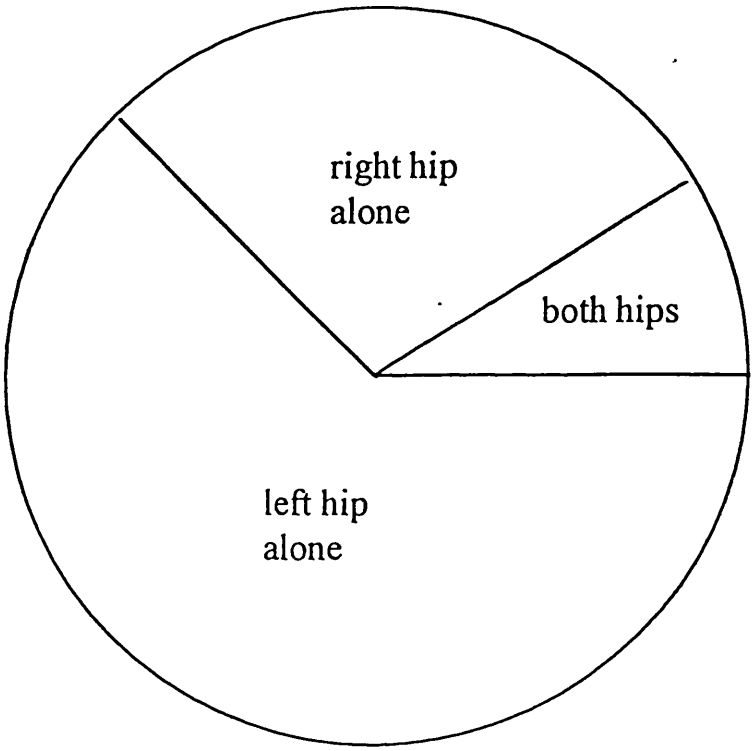
Clinical examination

On clinical examination, both hips were reported to be affected in 19 children, but 2 were found to have an affected left hip only and 1 an affected right hip only. Right-sided instability and no instability were reported in 2 and 7 children respectively who received surgery for the left hip and left-sided instability and no instability were reported in 1 and 6 children respectively who received surgery for the right hip.

Imaging

In children for whom only one hip was reported to be affected, ultrasound examination revealed both hips to be affected in 8 children, and no abnormality in 3 children, and X-rays showed both hips to be affected in 3 children and no abnormality in 4 children.

Figure 4.6 Cases of a first operative procedure: side affected (n=317)



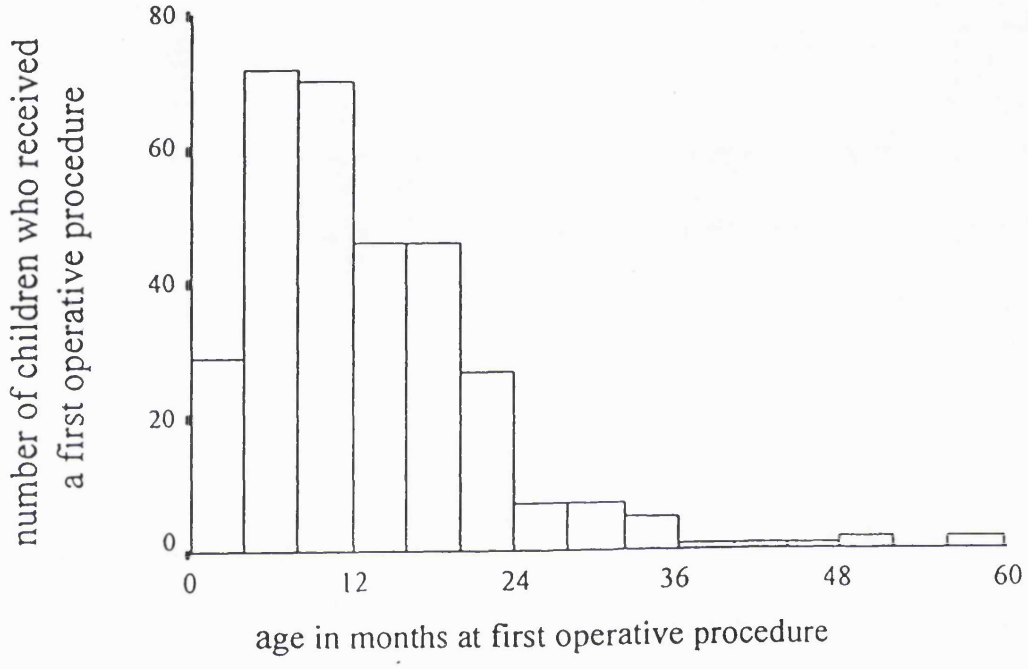
Mode of presentation

More than one third (37%) of children had been brought to the attention of the health service by their parents or other family member at a median (IQR) age of 15 months (12 to 18). Less than one third (32%) of children had been detected by screening or routine surveillance before the age of 3 months. A further 4% had been identified in the course of follow up for children at high risk of CDH, while 3% of children had been discovered incidentally while under medical care for another condition. The remainder (24%) were detected by routine surveillance after 3 months of age.

Age at first operative procedure

The median (IQR) age at first operative procedure was 11 months (6 to 17) and only 8% were detected after the age of 24 months (Figure 4.7). The median (IQR) interval between detection and surgery was 1 month (1 to 3).

Figure 4.7 Distribution of age at first operative procedure



Prior splinting

Prior treatment with an abduction splinting appliance had been given to 67 (21%) of children for a median (IQR) duration of 8 weeks (5 to 14). The most commonly used splint appliance was the Pavlik harness (62%). Other splints included the Promedic, the abduction brace, the Aberdeen, the Denis Browne and the Pyford. Among those previously splinted the median (IQR) age at time of first operative procedure was 6 months (4 to 11) and median (IQR) interval between detection and surgery was 6 months (3 to 9) compared to 12 months (8 to 18) and 2 months (1 to 4) respectively in those who had not received prior splint treatment. A period of skin traction may be undertaken before surgery, and there was a period of more than two months between removal of the splint and surgery for more than half (53%) of the children who received splinting.

Diagnostic tests

A first operative procedure may include a diagnostic test (for example, arthrogram). A pelvic X-ray was performed prior to surgery in 263 children (85%). Other than clinical examination, no tests before the first operative procedure were reported for 21 (7%) children but ultrasound was used to further assess the hips of 74 children (24%). The type of ultrasound performed was reported for 56 children: dynamic alone in 30%, static alone in 21% and both in 48%. Only 3 children had received primary screening with ultrasound.

Complexity of first operative procedure

The type of first operative procedure was grouped according to complexity (Table 4.13). The least complex intervention (arthrogram, examination under

anaesthesia, application of a plaster of Paris cast, or some combination of these) was the first operative procedure in the greatest number of children (43%). A pelvic osteotomy was the first operative procedure in only 6%. Only one child (1.5%) who had been previously splinted received a major plus or complex major first operative procedure compared to 26 children (10%) who had not been previously splinted (χ^2, p=0.02).

Table 4.13 Complexity of the first operative procedure

Type of first operative procedure	Description	Number of children
minor	arthrogram/EUA[a]/PoP[b]/closed reduction	138
intermediate	adductor tenotomy	104
major	open reduction	49
major plus	femoral osteotomy	8
complex major	pelvic osteotomy	19
TOTAL		318

[a]EUA examination under anaesthetic
[b]PoP application of a plaster of Paris cast

4.2.6 Children treated for CDH in the Republic of Ireland

One child who received a first operative procedure and 57 children who received abduction splinting in the Republic of Ireland were notified to the BPASU and OS schemes. This corresponds to a prevalence of a first operative procedure of 0.02 per 1000 live births and a prevalence of abduction splinting of 3.4 per 1000 live births (95% CI: 2.6, 4.3). The prevalence of abduction splinting is more than twice the prevalence obtained in this study for the UK, but direct comparison is limited due to potential under ascertainment in both countries.

Child who received a first operative procedure

This child was female, detected at 21 months of age as the result of parental concern. Her right hip alone was affected and she received an open reduction and innominate procedure (i.e. complex major procedure) as initial surgery. She had no risk factors for CDH.

Children who received abduction splinting

Nearly all (95%) of infants who received abduction splinting were detected by screening and the median (IQR) age at detection was 2 days (1 to 3). Of the remainder, one child presented clinically at five months and two children were detected at five and six months of age respectively through follow up programmes for children at high risk. Pelvic X-rays were taken of the latter three children only. Ultrasound imaging of the hips was not undertaken in any child.

Double nappies were the initial treatment for 10 children (18%), in two of whom this was followed by application of a formal splinting appliance. The Pavlik harness was the most frequently used splinting appliance (57% of children). The other background characteristics of the children are shown in Table 4.14.

Table 4.14 Background characteristics of children treated in the Republic of Ireland

Characteristic	
female: male ratio	4:1
median (IQR) birthweight (kg)	3.8 (3.2 to 4.1)
median (IQR) gestation (weeks)	40 (40 to 42)
ethnic group (%)	100% white
hip affected (%)	both (29); left only (43); right only (29)
firstborn (%)	44
risk factors (n)	16 (36%):
positive family history	8
breech presentation	8
talipes	1

These data are similar to those for the children reported from the UK.

4.3 VALIDATION

4.3.1 Validation of cases of abduction splinting

Replies were received to 22 (71%) requests for other sources of cases of abduction splinting. Less than half the area of England, Scotland and Wales is served by local congenital malformation registers.[230] Of the registers identified as including cases of CDH, none recorded whether and when a splinting appliance was applied. Reference to the case notes for all possible cases of abduction splinting would have been expensive. No register was identified for Wessex, and the register for the Northern region did not include CDH. The Greater Glasgow Health Board is reported to be the only health board in Scotland with a well-organised congenital malformations scheme.[231] A listing from this register identified 5 children who were less than a year old when they were diagnosed with CDH during the study period, and thus might have been splinted. All these children had been reported to the OS

scheme, 4 of whom had received surgery without prior splinting and one of whom had been splinted during the study period.

An independent audit of case reporting to the BPASU at the Duchess of York and Wythenshawe hospitals conducted during the period of the surveillance study found the OPCS coding system inadequate for identifying children treated with abduction splinting: a search of the computer system identified 18 children in one hospital and 54 in the other with a diagnosis of CDH. However, reference to the medical records revealed that only one child in the former hospital had been treated with abduction splinting. This child had already been notified to the BPASU scheme. No other validation of the confirmed cases of abduction splinting was possible.

4.3.2 Validation of cases of a first operative procedure: HES data

From the HES data, 51, 81 and 139 cases of a first operative procedure for CDH were identified in the Wessex region, Northern region and Scotland respectively. Reference to the medical records revealed that 35, 38 and 73 of these cases, respectively did not meet the study criteria. The most common reason for ineligibility was that a prior operative procedure had been undertaken (Table 4.15).

Table 4.15 Reasons for ineligibility of children identified by HES data

Reason for ineligibility	Number of children			
	Wessex	Northern	Scotland	Total
not first operative procedure	13	34	57	104
not CDH	8	0	3	11
child over 5 years	12	1	1	14
CDH but no surgery	2	1	6	9
dislocated hips secondary to another condition	0	2	6	8
TOTAL	35	38	73	146

4.3.3 Source by which children identified, by region

The number of children identified by HES data, by the OS scheme and by both sources is given in Table 4.16.

Table 4.16 Source by which children identified, by region

	both OS scheme and HES data	HES data alone	OS scheme alone	TOTAL
Wessex	11	5	7	23
Northern	14	29	2	45
Scotland	38	28	9	75
TOTAL	63	62	18	143

4.3.4 Effectiveness of search strategies

Forms were completed for 271 children, of whom less than half (46%) were eligible cases. The most common reason (71%) for ineligibility was due to the first operative procedure occurring before the 1st April 1993 or after 30th April 1994. This lack of specificity was expected in the Northern and Wessex regions where records are unlinked but poorer than expected in Scotland. An attempt was made to develop a more specific search strategy, using the largest list of cases, from Scotland. Exclusion of cases with a operative procedure code of X48 or X49 (i.e. without a diagnosis of CDH) reduced the list to 102 cases. Use of this search strategy would have missed 6 eligible cases who were not coded with a diagnosis of CDH. If this modified strategy were extended to include codes for 'other congenital deformities of the lower limb' and 'pain in joint' in association with 'application of cast', these six cases would have been identified at the cost of searching only 2 additional unnecessary sets of medical records. With hindsight, the 66 eligible Scottish episodes could have been confidently identified from a more specific search of 110 medical records, rather than 139. Although the cost of completing a few extra forms at a visited hospital, is marginal, the cost of an additional visit to a far flung location may be relatively high.

4.3.5 Ascertainment-adjusted estimate of the prevalence of a first operative procedure

Use of the formulae given in equations 3.1 and 3.2 (page 76) suggested that 17 (95% CI: 5, 30) cases were likely to not to have been identified by either the OS scheme or the HES data. The prevalence of a first operative procedure in the validated area was calculated for each source separately and for the combination of the data sources, after adjustment for case-under ascertainment (Table 4.17).

Table 4.17 Under ascertainment-adjusted prevalence of a first operative procedure, by source

Source	n	Births	Prevalence (per 1000 live births)[220,221]	95% confidence interval (per 1000 live births)
OS	81	152775	0.53	0.41, 0.65
HES	125	152775	0.82	0.67, 0.96
ascertainment-adjusted	160	152775	1.05	0.97, 1.13

The reasons for under ascertainment to the OS scheme cannot be established clearly from the validation study. However, it was possible to identify the treating consultant for cases ascertained from Scotland. This showed that of the 28 cases identified by HES data but not reported to the OS scheme, 22 (79%) were treated in a month during which the responsible consultant had not returned his reporting card to the OS scheme and suggests some association between poor card return rate and case under ascertainment.

The ascertainment-adjusted prevalence was calculated for each validated area separately (Table 4.18).

Table 4.18 Geographical variation in the ascertainment-adjusted prevalence of a first operative procedure

Area	n	Births	Prevalence (95% CI) (per 1000 live births)[220,221]
Scotland	81	68575	1.2 (1.1, 1.3)
Northern	49	41814	1.2 (1.0, 1.3)
Wessex	26	42385	0.6 (0.5, 0.7)

4.3.6 Comparison of the characteristics of cases by source of identification

The additional children identified by the HES data were examined to establish if they were substantially different from the children reported to the OS scheme (Table 4.19). Differences between proportions were tested using Fisher's exact test for dichotomous variables and the two sample unpaired t-test was used for continuous variables, using logged values where appropriate.

Table 4.19 Comparison of characteristics of children identified by Hospital Episode System data (HES) only and those ascertained through the Orthopaedic Surveillance Scheme (OS)

Characteristic	HES	OS	p-value of tests of significance between groups
n	62	81	
female (%)	82	93	0.07
mean birthweight (kg)	3.4	3.3	0.93
median gestation (weeks)	40	40	0.40
>1 risk factor (%)	44	38	0.61
breech presentation (%)	20	30	0.23
talipes (n)	3	2	0.65
positive family history (n)	7	9	1.00
median (IQR) age detected (months)	6 (0 to 13)	9 (2 to 15)	0.04
identified by screening or surveillance (%)	67	44	0.01
underwent prior abduction splinting (%)	27	15	0.92
median (IQR) duration of splinting (weeks)	11 (5 to 16)	8 (6 to 14)	0.49
underwent prior traction (%)	48	47	0.87
surgery for both hips (%)	11	6	0.22

IQR = interquartile range

No differences were significant at the 5% level for gender, birthweight, gestation, the side affected, or the proportion with risk factors (Table 4.19). The proportion who received prior traction, or abduction splinting, was similar in each case. The mean duration of splinting was also similar. However, children identified through HES data alone were more likely to be detected by screening or surveillance, to be

detected earlier and to receive a less complex procedure (Table 4.20), (χ^2 test for trend, p=0.01). These children were perhaps less memorable for the surgeons.

Table 4.20 Complexity of first operative procedure, by source

Complexity of first operative procedure		% cases identified by HES only	% cases reported to OS scheme
minor		58	38
intermediate		34	42
major		8	20
(3 divisions:)	major	5	14
	major plus	2	2
	complex major	1	4

HES = Hospital Episode System data
OS = Orthopaedic Surveillance

4.3.7 Variable catchability

Subgroup variation can be overcome by stratifying the population by probability of capture, deriving estimates for each of the strata, and summing these to derive an estimate for the total population.[232] It was decided to stratify by complexity of surgery. Capture-recapture analysis was performed separately for each group of surgical complexity (Table 4.20). This revealed that a further 3 cases were likely to have been missed and the estimate of the prevalence of a first operative procedure was revised to 1.06 (95% CI: 0.09, 1.2) per 1000 live births for the validation areas.

4.3.8 Application of results of the validation study to national figures

The capture-recapture analysis revealed that in the validated areas 50% of eligible cases were likely not to have been reported to the OS scheme (81/163). When this

result was applied to the UK figures, the corrected prevalence of a first operative procedure in the UK was 0.78 per 1000 live births (95% CI:0.72, 0.84), in England and Wales alone 0.66 per 1000 live births (95% CI: 0.60, 0.72) and for CDH not detected at the first or second screening examination 0.62 per 1000 live births (95% CI: 0.57, 0.68).

5: DISCUSSION

5.1 SURVEY OF SCREENING PRACTICES

5.1.1 Potential sources of bias

The high level of coverage achieved in this survey suggested that any bias due to non-response was likely to be small. Information requested was factual, rather than subjective and therefore the respondent and recall bias and bias associated with postal surveys owing to the respondent reading through the questionnaire before answering the first questions was likely to be low. The factual content may also have encouraged response since reduced response rates have been reported to surveys of attitudes and facts, rather than facts alone.[233] Sending a duplicate form with each reminder letter may have further encouraged response.[233]

Only one paediatrician was contacted for each unit and, in units lacking an agreed policy or where individual clinicians follow different policies, the responses elicited may not have represented the care received by all children born in that unit. Information regarding ultrasound and management was obtained from radiologists and orthopaedic surgeons in 45% and 57% of units respectively. In the remaining units, it is possible that the responses from paediatricians regarding ultrasound and management were less accurate. Information was collected on reported, rather than actual practices and for some units the responses may have reflected policy or 'best guesses', rather than actual practice. However, a survey of actual rather than reported practices would have required visits to each unit and direct observation, which would have been expensive, was not considered feasible or appropriate. Subsequently, data from this survey have informed the design of a multicentre

randomised trial of ultrasound imaging in the management of clinically-detected neonatal hip instability (NHI), coordinated by the Perinatal Trials Service in Oxford. Information from the survey was used to identify and enrol centres using ultrasound to this trial. No information subsequently verified in this manner was found to be inaccurate.

Although efforts were made to produce a simple form with clear instructions, and to pilot the form widely, some questions were answered less well than others and it is possible that some questions were misunderstood. For example, a potential difficulty with the questions regarding the identification and management of 'high risk' infants (questions 11-13; Appendix 3.1), was that respondents might have assumed that they were designed to ascertain their knowledge of risk factors for CDH, rather than whether infants at high risk of CDH were screened or managed differently to those infants not at risk. Internal consistency checks were included, for example, question 14, which asked whether the procedures described for high risk infants were carried out irrespective of clinical findings. This subsequently revealed that 35 respondents had incorrectly reported the existence of a high risk policy irrespective of clinical findings. The other important internal consistency check was to ensure that the replies to questions 15a,15b,19a, and 19b regarding ultrasound in management and question 12 regarding ultrasound in the screening of high risk infants were consistent with questions 7-10 concerning the use of ultrasound examination (Appendix 3.1). No inconsistencies were identified for these questions.

A few paediatricians supplied descriptions or protocols of their local practice in

addition to completing the forms but practice could always be satisfactorily described by the available tick boxes, except where management practices differed for irreducibly and reducibly dislocated hips. Given the wide variations in reported practice, particularly regarding the definition of a high risk infant and the management of clinically detected hip instability, it was difficult to design a questionnaire that allowed a respondent scope for open-ended response but ensured that specific questions were answered without a major interpreting and coding exercise. In retrospect, one change would be to consider treatment practices separately for irreducibly dislocated, reducibly dislocated, and dislocatable hips in turn but this issue did not emerge in the pretesting of the questionnaire.

5.1.2 Comparison to previously published data

There have been no previous national surveys of screening and management practices for CDH, other than one survey of health districts[234] which reported that in 1991, only 44% of health districts had a designated officer to oversee the screening process. In 50% of these districts this was an orthopaedic surgeon and in 9% a paediatrician. In the current study, which achieved a higher response rate (92% of maternity units compared to 70% of health districts), only 40% of units reported a designated officer. Paediatricians were more likely to be cited as responsible for keeping the programme under review in the current survey (32% of units) but this may reflect the specialty of the survey respondents. Co-ordination of both monitoring and service provision is complicated because paediatricians are primarily responsible for initial screening, orthopaedic surgeons for management and radiologists for the performing and reporting of ultrasound. Although no respondent identified a public health physician with overall responsibility for the

screening service, this may be the most appropriate specialty.

Divergence in current practice from the existing recommendations for the screening and management of CDH[1] reflects uncertainty regarding best practice as well as changes in maternity service provision subsequent to the latest recommendations made in 1986. A high proportion of units were unable to comply with the prescribed schedule regarding the second examination, probably owing to a trend to earlier discharge from hospital of birth. The value of a second examination may be in revealing previously unsuspected hip instability.[80] However, a second examination was attempted prior to discharge in only 8% of units, and was performed within a few days rather than ten days which may reduce its value.[80] As a consequence of recent changes in maternity services,[235] coverage for this examination may be further reduced, or be increasingly within the domain of primary care. If this is the case, additional training for primary care and community health professionals (who are reportedly less proficient at the O-B manoeuvre)[130] will be required. Although in one study[230] a single examination 24 hours before discharge from hospital of birth was found to yield a similar number of abnormalities as two neonatal examinations, the number of children in each group was too small to have detected anything but a large difference between the groups.

Since the current survey was based on maternity units, information regarding the recommended six week examination, which is carried out in primary care, was not obtained. The six week examination may be deferred in practice by 1-2 weeks as a result of the accelerated immunisation schedule, but whether this delay reduces the opportunity for conservative treatment of NHI to be effective is not clear. Timing

of the examination may also affect coverage as well as test performance, but information on this is lacking.

Training in clinical examination is of crucial importance since the O-B test is neither robust nor reliable (section 2.3.3). However, in this survey, primary clinical screening for neonatal hip instability was almost exclusively performed by junior paediatricians for whom formal training with a 'Baby Hippy' simulator model or a lecture or a video was provided in only two-fifths of units. Although NHI is considered a common malformation, few junior paediatricians will experience the feel of an unstable hip. Junior doctors cannot use real babies to train in the O-B manoeuvre as there is a view that this may provoke dislocation.[4,35,38,83,127,206] In addition, the 'Baby Hippy' does not adequately simulate the range of normality, and at the time of writing, is not easily available in the UK. The difficulty of ensuring adequate training of personnel who undertake screening for a limited period, has led to the view that there is a need for specifically trained personnel, such as physiotherapists, dedicated to hip screening.[84,86,236,36] This has implications for training midwives who have an increasing role in neonatal care.[235]

5.1.3 Performance and reporting of ultrasound imaging

There is limited consensus internationally in the performance and reporting of ultrasound examination of the hip. While static ultrasound examination is used in some European countries,[152,153] and dynamic examination in North America,[237] the findings of this survey indicate that, in the UK and Irish Republic, both methods are performed, either alone or in combination, with little consistency in the methods of reporting ultrasound appearances. Although interobserver agreement may be better

for the reporting, rather than the performing, of ultrasound imaging of the hips,[163] agreement in reporting, even among experienced professionals, has been found to be poor.[162] This may be of particular importance where the clinician responsible for management does not perform and report ultrasound images, which was the case in more than two-thirds of the units in this survey.

5.1.4 Management of infants with a presumptive positive screening test

Ultrasound imaging of the hip may have the potential, when used as a secondary screening test, to increase the specificity of the O-B test. However, the effectiveness of this policy has not been adequately evaluated. In one controlled trial[155] a 71% reduction in treatment was reported among infants with clinical hip instability allocated, on an alternate basis, to ultrasound compared to those allocated immediate treatment, but the trial was too small and the follow up period too short to assess reliably the outcome for those left untreated. The current survey highlighted the increasing use of this unevaluated practice, which in 1993 was undertaken in 65% of maternity units in the UK and Irish Republic. In view of this, a trial of ultrasound in the management of the clinically abnormal neonatal hip has been initiated, and is due to report in the year 2000.

There have been no randomised trials of different treatment regimes, although the effectiveness of early non-surgical treatment is central to the rationale for screening, and the optimal timing, nature and duration of treatment of infants with hip instability is unclear. The proportion of infants treated is strongly influenced by the age at which diagnosis is confirmed and treatment started, as hip instability, when detected within the first days of life, may resolve spontaneously[29,38,79] by the time

of a subsequent assessment. In only 2 units were children treated on the basis of one positive clinical examination, while in only a third and a quarter of units was a policy of 'immediate treatment' reported for dislocated and dislocatable hips respectively.

The importance of follow up of infants with spontaneously resolving NHI has been emphasised, although both the age at which good outcome is assured, and a reliable means of assessing outcome are uncertain. Continued follow up of such infants was reportedly undertaken in 90% of units, with half of these units performing an X-ray at 3-6 months. Age at discharge from follow up ranged from 3 months to weight bearing age.

Centres varied markedly in their reported treatment of irreducibly dislocated hips, with some centres initiating conservative treatment, usually with a Pavlik harness, to encourage the hips to reduce without surgery and yet others offering a closed reduction and plaster of Paris cast at a few months of age as their first line treatment. Evidence to support either view is lacking, although in one trial in which the effectiveness of a hip spica plaster and a dynamic plastic harness were compared, there was no difference in outcome or in the rate of complications between the two groups.[98]

In the absence of trial evidence, potential differences in outcome from the use of different splint appliances are not known, and as many as fifteen different splinting appliances were used in the UK and Irish Republic. The duration of splinting, which was often reported as a range, varied markedly between units, reflecting

clinical uncertainty. Ultrasound may have a role in determining treatment duration, as it can be used to image the hips to monitor progress and ensure a concentric reduction with some splint appliances in situ, while avoiding the hazards of repeated pelvic X-rays. In the current survey, 67% of those units with ultrasound used it to monitor progress and 38% to determine treatment duration.

Whether double nappies encourage an unstable hip to develop normally is unclear but few centres reported their use as a definitive treatment for neonatal hip instability. Their value in the interim before a diagnosis is confirmed may be to reassure parents that their child's hips are under medical care, and to encourage parents to attend further clinics, as much as to abduct the hips.

5.1.5 Identification of infants at high risk of CDH

It is recommended that children at high risk of CDH are followed up until weight bearing age,[1] specifically those with a family history of the condition, or with breech presentation, other congenital postural deformities such as talipes, birth by caesarean section, oligohydramnios, or fetal growth retardation. Only half of maternity units provided specific assessment in addition to routine clinical screening for such children. Not all units with access to ultrasound imaging reported following a high risk policy but all units with a high risk policy and access to ultrasound used ultrasound to further assess babies at high risk. Further assessment of children in units without ultrasound was by repeated clinical examination. A positive family history, which may not always be evident, or consistently ascertained by junior medical staff, was the only criterion in all units' high risk policy.

Although ultrasound imaging of the hips of high risk children whose hips are clinically normal is currently undertaken in 36% of maternity units, this practice has not been fully evaluated in a randomised controlled trial. The detection of children who are clinically normal but have mildly dysplastic acetabulae may result in additional children receiving treatment for whom the benefits are unclear. In one study in the UK, the prevalence of late-presenting CDH was not reduced following the introduction of selective ultrasound screening.[137] In the trial of delayed splintage[155] described earlier, 15% of a third group of matched control children with clinically normal but ultrasonographically abnormal hips at birth were subsequently found to be normal, without treatment, by 6 months of age. The cost-effectiveness of selective primary ultrasound screening for children at high risk has been examined in a quasi-randomised trial in Norway.[151] Although the prevalence of late-presenting CDH was not reduced among those screening by ultrasound, the period of follow up was relatively short. It was concluded from this study that all girls, but only boys at high risk (including those with NHI) should receive primart screening with ultrasound. Furthermore, the prevalence of abduction splinting in each arm of the Norwegian trial was much higher than that reported in similar studies in the UK. Evidence to support the introduction of this policy in the UK is lacking.

5.2 SURVEILLANCE STUDY

5.2.1 Sources of bias

5.2.1.1 Completeness of the OS and BPASU reporting bases

If, at the outset, membership of the OS scheme had included only members of the BOA, the largest professional organisation, 12% of relevant surgeons would

have been missed. Comparison to the manpower census data suggested that, by using multiple sources of respondent ascertainment and monitoring new appointments, only 3% of surgeons had not been included in the OS reporting base. For the BPASU, the coverage of the reporting base appeared to be lower, with 8% of eligible paediatricians not included in the BPASU. This may reflect differences between the schemes in the methods used to compile and maintain the reporting base. However, this is unlikely to have reduced case ascertainment in this study since few paediatricians who were not included in the BPASU treated CDH during the study period. The BPASU and the BOA had assumed a higher proportion of consultant paediatricians and orthopaedic surgeons respectively to be members of their professional organisations, although it had been expected that paediatricians not included in the BPASU were likely to be community paediatricians whom historically may have been less associated with the BPA than hospital-based paediatricians.

This exercise improved the completeness of the reporting base of the BPASU, and established a method for routine audit which will be used by the BPASU in future.[238] However, the results indicate that the effect of incomplete reporting bases for the OS and BPASU schemes respectively was likely to be small for the current study.

5.2.1.2 Return of cards and forms

One measure of compliance with a surveillance scheme is the proportion of reporting cards returned. This may be described as the *response rate*, but could perhaps be more accurately termed the *card return rate*, since a completed card

does not guarantee a complete response in terms of the cases ascertained. The effect of including in a surveillance scheme those clinicians who never, or only rarely, see a child with a notifiable condition may be in two directions: clinicians may have little interest in the study and fail to make nil returns; alternatively, they may be motivated to participate, without the disincentive of requests for further information consequent on reporting a case. The former may reduce, and the latter inflate, apparent compliance without affecting case ascertainment. In either case, the card return rate may be a misleading measure of the success of the reporting scheme. Further, while it is possible to calculate the proportion of participating clinicians who make a monthly return, there remains the question of how many treating clinicians have not been included in the scheme. The OS scheme card return rate was good, and although not as high as that for the BPASU, was higher than that of the dermatologists, pathologists and rheumatologists reporting to the BPASU, who achieved rates of 62%, 63% and 63% respectively.[207] This suggests that it is possible to achieve good compliance with a reporting scheme in a relatively short period (less than three months preparation) among specialists with no prior experience of an active reporting scheme. Publicising the OS scheme among orthopaedic surgeons, providing feedback, and regular reminders are likely to have improved response.

The initial questionnaire employed in the OS scheme allowed 708 surgeons who did not ever treat children to be excluded, thus minimising the workload of surgeons as a whole and saving resources. Both paediatricians and surgeons were informed that the other specialty was simultaneously requested to report cases of abduction splinting and in some cases, arrangements were made for one

or the other specialty to undertake this on behalf of both. This did not wholly eliminate duplicate reporting, however.

The implications of non-response vary with the aims of the study, as well as with the response rate. When the aim is to identify a small proportion of the population with a particular characteristic, as here, there is the theoretical possibility that all the respondents who did not participate treated cases with that characteristic. For example, for a rare condition, the impact of non-respondents might be considerable despite a high response rate of say 95%, if all 5% of the non-responders had seen and treated cases.[172]

Green and yellow forms were completed for 72% and 78% of notified cases of abduction splinting and a first operative procedure respectively and 75% and 76% of notified cases for which forms were returned were eligible for inclusion. The most common reason for ineligibility (39% of ineligible cases) was the initiation of treatment outside the study dates. This may be due to the completion of a form in respect of a child other than the child originally notified or erroneous notification of a child treated in a different month. Notifying clinicians frequently requested the researcher to provide names for the children for whom follow up forms were outstanding, although the initial notification included no such details. Failure to return the follow up form may have reflected failure to recollect the name of the child notified. In future studies of common conditions, clinicians should be asked to provide identifying details of children at notification, subject to ethical approval. This may require the reporting cards to be returned in a sealed envelope rather than as a postcard to preserve

confidentiality. The OS scheme was modelled on the BPASU which had reported success among paediatricians in the UK and Irish Republic and its clones in other countries. However, the BPASU was established as a unified approach to obtaining information on several rare conditions, rather than one relatively common condition from a targeted group of clinicians.

Case eligibility was carefully checked from the information requested on the follow up forms to ensure that the prevalence of treatment was not overestimated. Comparison of the background information for cases reported to the OS scheme and identified by HES data, and for 9 children reported to the OS scheme both as cases of abduction splinting and as cases of a first operative procedure indicated that the information provided by clinicians was accurate. Surgeons had difficulty in providing information on birthweight and gestation but this was generally successfully obtained from the child's general practitioner. Data for each variable were at least 80% complete. It had been expected that the reported test findings would have included information on the type of abnormality, i.e. whether the hip(s) were reducible or irreducible, unstable, or stable, but usually the description "CDH" was given. With hindsight, this information could have been requested explicitly, for example, by tick boxes, but pretesting the forms did not reveal this to be a problem.

5.2.1.3 Under ascertainment of cases

The completeness of the reporting bases, the card return rate and the form return rate were measurable but failure to report a case could not be determined without multiple sources of case ascertainment. Capture recapture analysis provided an

opportunity to estimate the number of first operative cases not identified by HES data or reported to the OS scheme. Case ascertainment was found to be only 50% for the OS scheme, despite a reporting base which was 97% complete, a mean (median) card return rate of 70% (92%) and a form return rate of 76%. A second source of case ascertainment in which case definition was comparable to the first source was very important to the validity of the study.

Capture-recapture analysis required that the lists were closed, that all and only true matches could be identified, that the sources were independent and that each case had an equal chance of inclusion in each list. The first two assumptions were clearly met: neither the HES data nor the OS scheme were open to change and individuals were easily identified and matched between the two sources. While the reporting clinician was responsible for recording the diagnosis of CDH in the medical records, and the medical records formed the basis of the coding for HES data, the processes by which an individual was included in HES data or notified to the OS scheme were independent. However, in investigating the possibility of variable catchability, it was found that the cases not reported to the OS scheme tended to have received less complex surgery, and the ascertainment-adjustment was corrected accordingly.

The most recent annual publication of the BPASU (now the Surveillance Unit of the Royal College of Paediatrics and Child Health) reported that recent BPASU studies have used multiple sources of case ascertainment and that case ascertainment by the BPASU scheme is on average 80-90%.[238] Capture-recapture methodology has been used in a variety of epidemiological studies but its first use in a BPASU-related study was to estimate the prevalence of insulin-

dependent diabetes.[205] Comparable lists were requested simultaneously from specialist diabetes nurses and health visitors, and HES data were obtained for 12 NHS health regions. However, confirmation of cases was sought by repeat contact with paediatricians rather than by reference to medical records, and was not received for 164 cases (30%), including 36 cases originally reported by paediatricians. Thus nearly one third of potential cases could not be confirmed. Although potentially cheaper, this approach was not considered appropriate for the CDH study. In addition, it would have imposed a further burden on orthopaedic surgeons, already too busy to return forms in the first instance.

Children who received less complex (and potentially less memorable) surgery were less likely to be reported to the OS scheme. Ascertainment of cases of abduction splinting may have been affected similarly but this cannot be quantified due to the lack of a second independent data source. However, where comparison to independently compiled lists of cases of CDH was possible in Manchester and Greater Glasgow, no additional cases were found that had not been reported to the BPASU or OS schemes.

5.2.2 Comparison to the published literature on prevalence of a first operative procedure

Before screening was introduced

In the current study, the ascertainment-adjusted prevalence of a first operative procedure for CDH in the UK was 0.78 per 1000 live births (95% CI: 0.72, 0.84) and, in England and Wales alone, 0.66 per 1000 live births (95% CI: 0.60, 0.72). These estimates both fall within the range of the prevalence reported in unscreened

populations (0.66 to 1.5 and 0.66 to 0.91 per 1000 live births, respectively). Use of a historical comparison is not ideal and there are a number of possible explanations for this similarity. A correction for under ascertainment of cases was made to the current data, and case ascertainment may not have been complete in the unscreened populations. It is possible that the true prevalence before screening was higher than estimated, or that the underlying prevalence has risen, even though screening has effectively detected and averted a proportion of the cases of CDH. Alternatively, the current screening programme may have had little or no effect on the number of children developing established CDH.

Knox and his colleagues, who have undertaken careful and important work in this area, compared estimates of the prevalence of CDH pre and post screening using multiple sources of data from Birmingham.[62] They calculated the prevalence of CDH in 1950-54 (without screening) to be 0.91 per 1000 live births compared to the period 1974-83 (with screening) from which the estimated prevalence of 'true CDH' was calculated to be 0.67 per 1000 live births. However, in the intervening 26 years, the proportion of the population of Caucasian origin had fallen from nearly 100% to 71%. They concluded that the prevalence of CDH requiring prolonged treatment (more than 17 weeks of abduction splinting) or surgery was no less in the Caucasian population in the period 1974-83 than it was before neonatal screening was introduced.

After screening was introduced

In the current study, the ascertainment-adjusted prevalence of a first operative procedure for children who were not detected by screening at the first or second

neonatal examinations was 0.62 per 1000 live births (95% CI: 0.57, 0.68) which is consistent with the median prevalence of 0.62 per 1000 live births (95% CI: 0.50, 0.83) reported from previously published studies (section 2.3.5). The wider confidence intervals of the estimate based on published data reflect the large variation in reported prevalence.

Estimates of prevalence in smaller populations are less precise than in larger populations, but in the current study, the validation exercise and capture recapture analysis allowed more accurate estimates of the prevalence of a first operative procedure to be obtained for Scotland and the Northern and Wessex regions. Table 5.1 compares the prevalence estimates of the current study with those of the most recently published figures from these geographical areas. The estimates obtained for the current study are slightly, but not significantly, higher, from those reported previously. In the current study, 11% of children overall were treated outside their region of birth and the proportion treated outside their district of birth may well be higher. Thus, follow up to ascertain false negatives based on a single district or single reporting centre may underestimate the number of children presenting late unless care is taken to ascertain the cases treated outside the district or region of interest.

Table 5.1 Estimates of prevalence of CDH after screening was introduced: comparison to the literature by relevant region or county

Place	Prevalence per 1000 live births (95% CI) Most recently published study	Period of cohort	Current study 1993-4
Scotland	1.1 (1.0, 1.2)[77]	1986-91	1.2 (1.1,1.3)
Wessex RHA	0.4 (0.2, 0.6)[141]	1988-92	0.5 (0.6, 0.7)
Northern RHA	0.9 (0.5, 1.2)[a]	1972-76	1.2 (1.0, 1.3)

[a] Dr Edmund Hey, personal communication

From the data in Table 5.1 it is possible that there is some geographical variation in the prevalence of CDH. A consistently higher prevalence of a first operative procedure has been reported from Scotland and Northern Ireland than in the remainder of the UK[68,142] (chapter 2.4.4). In the current study, higher rates were found in Scotland and the Northern region, compared to Wessex. These consistent differences may reflect the genetic composition of the local populations, but other factors such as colder weather which may encourage the tighter swaddling of infants, or diet, may be relevant.

In the current national study, 30% of the children who received a first operative procedure had previously been treated with abduction splinting. This percentage varied between regions from 0 to 40% and similar variation is seen in the published reports from individual centres.[63]

5.2.3 Characteristics of infants and children who received a first operative procedure in the current study

About a third (32%) of children undergoing a first operative procedure had been detected by screening or routine surveillance before the age of three months, and a further 4% were identified in the course of follow up for children at high risk of CDH. However, the largest proportion (37%) were not detected by screening but were brought to the attention of the health service by their parents, while a further third were detected by routine surveillance after 3 months of age. This emphasises the limited sensitivity of the current clinically-based screening programme and highlights the value of vigilant observation of babies' hips after the neonatal period for signs of limited abduction, unequal thigh creases, and unequal leg length, as well as the importance of parental concerns and the role of community health professionals.

Female and White children were found be at increased risk of CDH in the current study, which is consistent with previous studies.[21,22,239,240] The suggestion that premature babies who spend their first few days in a special care baby unit may miss routine screening and constitute a relatively high proportion of the children who develop CDH[85] was however, not supported by the findings of this study. In addition, no seasonal trend in month of birth was found, as has been previously suggested, but the effect of a seasonal influence may be less pronounced in children diagnosed after the neonatal period.[21] As in other studies,[15,21,228] the left hip was more frequently affected than the right and in two-thirds of children, the left hip alone was affected. Breech presentation at delivery or in the last trimester was the most commonly reported risk factor (15% of infants) which is comparable to other

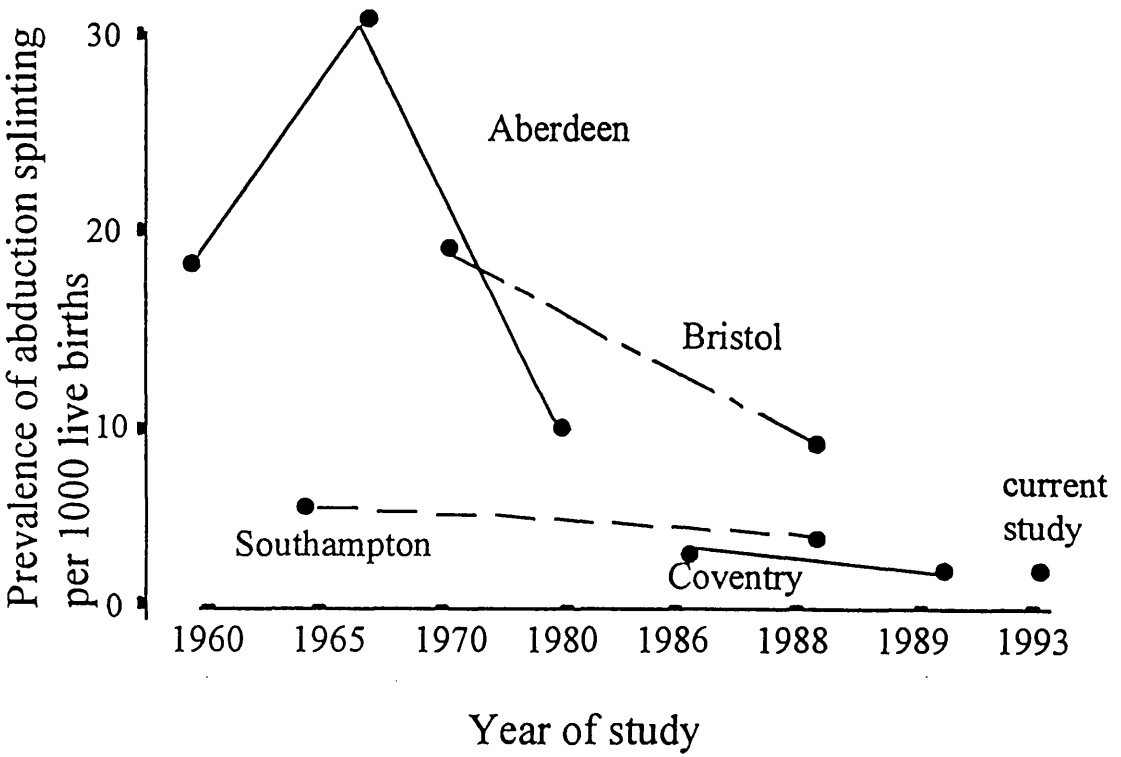
studies which for example, have reported breech presentation in 13% and 18% of children affected with CDH.[21,228] Routine data on the percentage of births nationally with breech presentation are not available (Dr Marion Hall, personal communication). The prevalence of a positive family history (11% of infants) was higher than has been previously reported.[241] Wynne-Davies[21] reported a prevalence of 6.5% in relatives of affected children from a study in which the parents of children with CDH were themselves examined and details of family history were likely to be well reported.

Although there is a lack of information regarding its predictive value, a high risk approach may allow a group of children at increased risk of CDH to be identified in whom CDH may be detected at an early stage through increased surveillance. However, among those receiving a first operative procedure and who were missed by screening, one or more risk factors for CDH were reported in only 30%, and a positive family history was reported in only 13%, and ascertainment of risk factors is likely to be good in these children. This suggests that focusing attention on children at higher risk of CDH will identify only a limited proportion of late-presenting children at an earlier age. In addition, affected children detected earlier as the result of a high risk programme would benefit only if treatment is therefore less invasive and/or long term outcome is improved. In the current study, children who received a first operative procedure after detection by screening were more likely to have a breech presentation, than those not detected by screening (24% and 11% respectively). This may be because infants with breech presentation are more likely to be O-B positive, or because the threshold for conservative treatment or the effectiveness of treatment is lower in these infants.

5.2.4 Complexity of surgery

'Open' procedures (direct surgery to the hip joint) are usually distinguished from 'closed' procedures (manipulative reduction to the hip joint) surgery. The former are more invasive and may be associated with a poorer prognosis, but the choice of open surgery may reflect a more severe condition. Alternatively, a closed procedure may be attempted in the first instance, or a first procedure may be purely diagnostic, for example, an arthrogram alone. Children with CDH usually receive more than one operative procedure and thus the complexity of the initial procedure for CDH may not reflect the severity of the underlying condition. Although nearly half of all children in the current study received a minor procedure initially, this gives no indication of the severity of the abnormality or the eventual outcome. A longitudinal study of the cohort would be needed to investigate further the complexity of the first operative procedure received, and its relation to the severity of the underlying condition, and outcome.

Figure 5.1 Secular trends in the prevalence of abduction splinting in four UK centres, 1960-1989

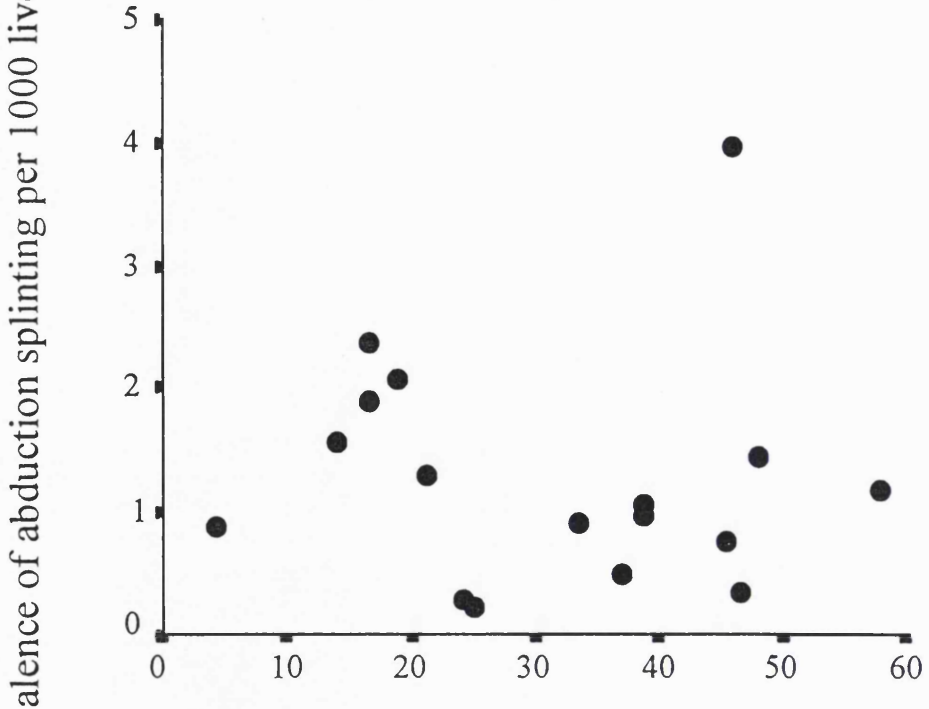


5.2.5 Prevalence of abduction splinting: comparison to the published literature

In the current study, the prevalence of abduction splinting was 1.6 per 1000 live births (95% CI: 1.4, 1.7), substantially lower than that reported previously.[63] Although no additional cases were identified in the two areas where a second source of cases was available, the populations at risk were small. Thus under reporting remains a possibility. The prevalence of abduction splinting may however have fallen over time with the increasing trend to confirm and treat NHI in those infants in whom instability persists beyond the first few days of life. Serial reports of the prevalence of splinting have been published from four UK centres. When plotted by calendar year (Figure 5.1), it would appear that the prevalence of abduction splinting has reduced within these centres over time.

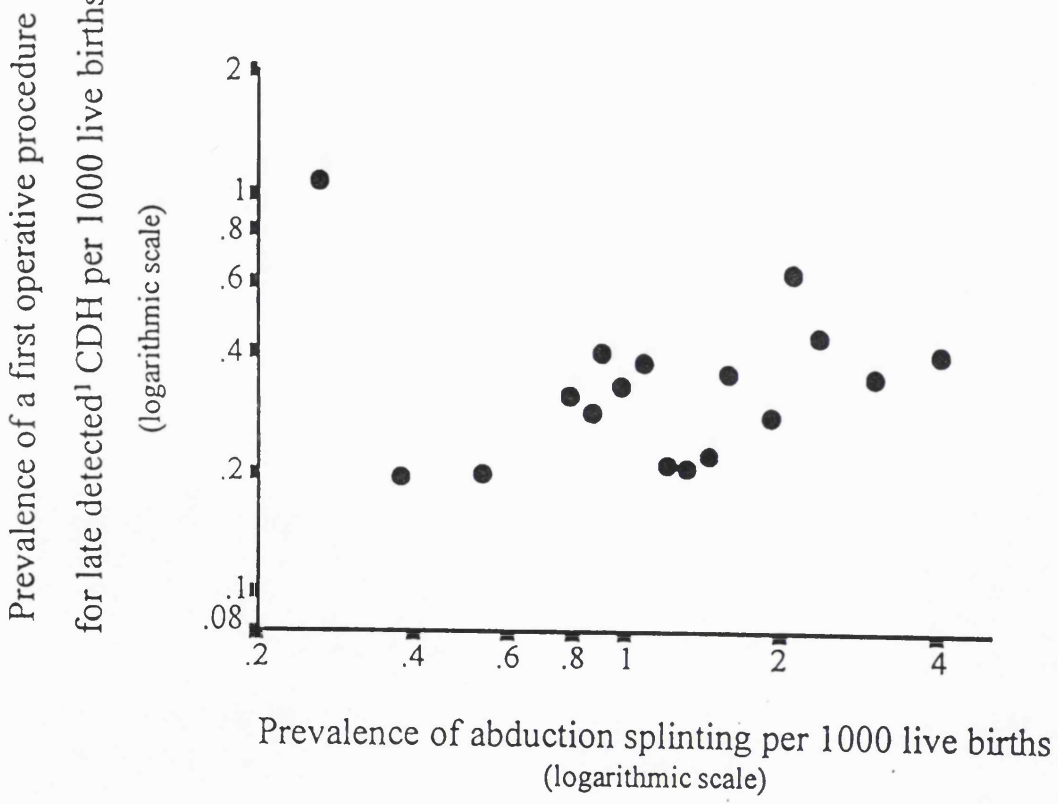
Differences in the performing and interpretation of the findings of the O-B test have been invoked to explain observed geographical variation in the prevalence of abduction splinting[236] but these may also reflect differences in timing of the O-B test. In the current study, no association was found between the regional prevalence of abduction splinting and the proportion of births for which a policy of immediate treatment was reported (Figure 5.2).

Figure 5.2 Prevalence of abduction splinting in relation to reported policy of immediate treatment of neonatal hip instability, by region



The specificity of the O-B test cannot be directly estimated because there is no diagnostic 'gold standard'. However, the prevalence of abduction splinting, despite potential under reporting, was approximately twice the reported prevalence of CDH in the absence of screening. Sensitivity and specificity are thought to be inversely related (section 2.1), which has been referred to as the 'borderline effect'. If this were the case, the prevalence of a first operative procedure in children not detected by screening would be expected to be inversely realated to the prevalence of abduction splinting. However, a lack of correlation or a positive correlation between the prevalence of splinting and the prevalence of late diagnoses has been reported.[44,62,63] In this study, no evidence of a relationship between the prevalence of a first operative procedure and the prevalence of abduction splinting was found (Figure 5.3), which suggests that neither the sensitivity nor the specificity of the O-B test are high. NHI may not in fact be the preclinical phase of CDH but a separate milder condition in the spectrum of hip disorders which shares similar characteristics with CDH. This theory has been considered using data from Birmingham for the period 1974-83.[62] As in the current study, children with late detected CDH were found to share similar risk factors with children with NHI, but to exhibit these risk factors to a lesser extent. The authors concluded that NHI and CDH 'appeared to be elements of a continuum'.

Figure 5.3 Prevalence of abduction splinting and of a first operative procedure for late-detected[1] CDH, by region



[1]not detected by screening

5.2.6 Characteristics of reported cases of abduction splinting

Among the infants treated with abduction splinting, there was an excess of female and White infants and a tendency to higher birthweight. A risk factor for CDH was reported in nearly half of infants (48%), of which breech presentation at delivery or the last trimester was the most common (23% of infants). Breech presentation and a positive family history were more commonly reported than in previous studies.[21,242]

6: CONCLUSION

6.1 Summary of findings

This first national study of screening and management practices for CDH revealed a five-fold increase in the use of ultrasound imaging of the infant hip over the last decade. In 1994, this technique was used in 69% of maternity units as a secondary screening test for infants considered to be at high risk of CDH or those with clinical hip instability. Hip ultrasound was rarely used as a primary screening test with universal primary screening by clinical examination being undertaken mainly by junior paediatricians. Only 50% of units had a policy for the identification and management of infants at high risk of CDH. The management of clinically detected neonatal hip abnormalities varied widely. The service was multidisciplinary, involving paediatricians, orthopaedic surgeons, radiologists, and radiographers. A single individual responsible for keeping the programme under review was identified in only 40% of units.

The high return rate of questionnaires and the completeness of responses achieved in this study of reported practice have allowed detailed and full information to be obtained about the screening and management practices for CDH in the UK and Republic of Ireland. In this survey clinical uncertainty regarding best practice was identified, as reflected in the departure from the current screening guidelines and the wide variation in management practices. From the survey, it appeared that the window of opportunity for a trial of ultrasound as a secondary screening test was closing. Information collected through the survey allowed a trial to be established quickly by identifying appropriate centres for recruitment.

The findings of the surveillance study showed that the prevalence of a first operative procedure was similar to that before screening was introduced. However, active surveillance identified only half of the children who underwent a first operative procedure. The validation exercise which employed a second data source and capture-recapture analysis allowed adjustment for the under ascertainment of operative cases and hence increased the validity of the study findings. The prevalence of abduction splinting may also have been underestimated but was double that of a first operative procedure. More than one third of those receiving a first operative procedure were children who were first brought to the attention of the health services by their parents, while just under one third had been unsuccessfully treated with prior abduction splinting. No risk factor for CDH was identified in two-thirds of those receiving a first operative procedure, but at least one risk factor was reported in nearly half of the confirmed cases of abduction splinting.

6.2 Implications for the current screening programme

The findings of this first national study revealed that the prevalence of surgery for CDH was consistent with the prevalence of CDH before screening was introduced. Although this may reflect alterations in the background prevalence of CDH as much as poor sensitivity of the screening programme, it nonetheless raises serious concerns about the ability of the current programme to detect children early. An effective CDH screening programme requires children to benefit from screening through earlier detection, successful and less invasive treatment of abnormal hips, and better hip function and development than would be the case in the absence of screening. Local audits have attempted to measure the effectiveness of screening for CDH using the prevalence of 'late' CDH, variously defined, or the prevalence of surgery, as a proxy

for outcome. The difficulties in identifying all members of a cohort who receive surgery have been discussed but additionally there remains the question of a suitable outcome measure. While surgical treatment may be less invasive as the result of earlier detection, complexity of initial surgery may not reflect the severity of the underlying condition, and hence is a better measure of process than outcome. One limitation of the current study is that it has provided information about the complexity of initial treatment but not the severity of the underlying condition. Follow up of children receiving surgery in Scotland and Wessex and the Northern region is proposed to enable presenting severity and radiological and functional outcome of this unselected and nationally representative sample to be identified.

Although information on disease severity would be useful, this was not considered in Leck's[147] estimation of the performance of the screening programme (section 2.3.5). He assumed a midpoint estimate of the underlying prevalence of CDH in northern European populations of 1.2 per 1000 live births (range 0.8 to 1.6) and used data from a study from which the estimated prevalence of late CDH was 0.21 to calculate a sensitivity of 83%. However, the data from the current study suggested a higher prevalence of late CDH nationally (0.62 per 1000 live births (95% CI: 0.57, 0.68)) and additional previously published data from the UK alone not included in Leck' figures (section 2.3.1) indicates a lower median prevalence of CDH before screening was introduced of 0.85 per 1000 live births (range: 0.66 to 1.5). Using these data and Leck's method, the sensitivity of the programme (27%) is much poorer than that reported by Leck, despite the use of a more conservative figure for the underlying prevalence of CDH. In addition, although the estimate of the national prevalence of abduction splinting from the current study is likely to be an underestimate, the odds

of being affected given a positive test result is 1:6, i.e for every affected child correctly identified, six are treated unnecessarily. This compares to 1:4.5 calculated by Leck.

There are a number of stages in the screening process which may be implicated in the disappointing performance of the screening programme. These include: the expertise of those responsible for undertaking the screening test, the screening test performance in relation to the natural history of CDH, and the effectiveness of early treatment. If the O-B test, when perfectly executed, is a valid and highly predictive measure of CDH, then efforts should be directed at improved training for existing or alternative (such as physiotherapists) screening personnel. However, there are some practical difficulties in assessing the performance of screeners for a condition such as CDH which is relatively rare and for which a confirmatory diagnostic test is lacking. While it seems undesirable to predicate a screening programme on the less experienced paediatric medical staff, direct evidence to implicate their lack of expertise as a cause of poor programme performance is lacking. An alternative explanation may lie in the natural history of CDH and its relation to NHI which is not fully understood. Hip instability may not, however, be the preclinical phase of CDH but perhaps a separate related condition. There is only indirect evidence to link CDH with NHI. Although Knox concluded that they were part of the same spectrum of disorders, observational studies to assess the predictive value of NHI for CDH have not be undertaken. If the O-B test for NHI is not a useful predictor of CDH, then alternative approaches should be evaluated in a randomised trial: these would include primary screening with ultrasound, as currently practised in Germany and Austria, as well as the option of no screening. The poor positive predictive value of the O-B test suggests that a significant number are receiving unnecessary treatment. This might be viewed as

acceptable were early treatment highly effective and free from adverse consequences, but there is little evidence to suggest that abduction splinting fulfils either of these assumptions. In the current study, infants treated with abduction splinting comprised 30% of the children requiring surgery, which raises concerns about its effectiveness. Abduction splinting has not been evaluated in a randomised controlled trial.

The role of ultrasound may be to detect infants at risk of subsequent dislocation or acetabular dysplasia in whom NHI is not a feature, thereby improving the sensitivity of the programme. Since the O-B test cannot detect acetabular dysplasia in the absence of NHI, it has been suggested that many of the abnormal hips that have been missed on clinical examination would not be detectable at birth without ultrasound imaging.[243] However, data from Germany and Austria suggests that the specificity of primary screening with ultrasound is poor when undertaken at birth. Furthermore, the predictive value of a dysplastic acetabulum in the neonatal period is unknown: acetabulae described as abnormal at birth have been reported as normal at five months of age without treatment.[155] Evidence from Norway, Germany and Austria suggests that there is considerable potential for over treatment with primary ultrasound screening.[81,152,153] This is not inevitable, since a much lower prevalence of abduction splinting was reported in the only published UK study of universal primary ultrasound screening, in Coventry.[146] However, the relatively low UK figure may have been achieved at the cost of intensive follow up, with 6% of all births followed for initially ambiguous ultrasound appearances and up to 8 scans per child. Ultrasound test performance outside the neonatal period is not known, although in Germany, a second universal screening examination at 4-5 weeks has just been introduced (Professor Rudiger von Kries, personal communication).

Evidence from the current study shows that in the UK and Irish Republic, ultrasound is used primarily as a secondary screening test. This increasing, but currently unevaluated, use of ultrasound imaging may have the potential to identify the false positives and thus improve the specificity of the O-B test. The Medical Research Council Working Party elected to evaluate this 'diagnostic' use of ultrasound before attempting a trial of primary screening methods because the window of opportunity to mount such a trial was closing, and it was considered sensible to address the value of ultrasound in high risk cases first. The MRC 'Hip Trial' is due to report in the year 2000.

The question of the most appropriate form of primary screening remains unresolved. Evaluation of the screening programme should clearly have preceded its implementation, and the strongest level evidence would be obtained from a randomised controlled trial. For a potential trial of primary screening methods, certain key issues relating to trial design would need to be addressed, including the specific approaches to be compared. Screening for CDH has formed an established part of clinical paediatric practice for more than a quarter of a century and the feasibility and acceptability of a trial will need to be explored. The costs of a large multicentre trial and the requirements for local ethical approval[244] have deterred doctors from entering their patients into trials[245] and a trial of screening methods for CDH would involve large numbers of babies and expense, in order to obtain sufficient statistical power to detect differences between groups. The acceptability to both clinicians and parents of a no screening trial arm is uncertain. The medico-legal implications would have to be addressed: 'missed' CDH is a major cause of litigation against paediatricians in the United States of America. In addition, although in the current study no risk factor was

reported for 69% of the late-detected children who received surgery, there may be ethical objections to the randomisation of high risk infants. Since nearly half the children with NHI in this study were reported to have at least one risk factor for CDH, a trial of primary screening methods excluding high risk children would take much longer, involve more babies, more expense and be less generalisable than one in which all babies were randomised. The choice of outcome measure(s), the optimal treatment strategy, and the standardisation of and training in the screening tests employed must be further addressed. A strong level of evidence would be required to persuade clinicians to discontinue the practice of a quarter of a century. Equally, strong evidence would be required to justify universal ultrasound screening since its introduction is likely to be expensive in both human and financial terms in relation to the potential benefits. A decision analysis would help to establish which specific research questions should be addressed, and hence the most appropriate trial design. There have been two published decision analyses of CDH screening with ultrasound to date, but these have been based on data from non-randomly selected case series from centres of excellence. These decision analyses did not evaluate all options and aspects of potential screening programmes (for example, the effect of the timing of examinations), and were heavily dependent on the relative costs of children detected and not detected by screening, and on the estimated prevalence of surgery for CDH in both screened and unscreened populations. However, cost data are limited and, as shown in this study, the variation in the estimated prevalence of surgery is considerable. A systematic review of the literature is required to ensure the derivation of parameters for the decision analysis considers all available evidence. Appropriate cost data will be available from the MRC Hip Trial. A trial in which children with a positive O-B test are randomised to receive abduction splinting or no intervention may

not be acceptable, but it may be possible to compare the effectiveness of conservative versus surgical treatment.

6.3 Implications for potential routine surveillance of the screening programme

The performance of the current national screening programme is not routinely monitored on a national basis and its value remains uncertain. Local audits have reached conflicting conclusions regarding programme effectiveness and have been hindered by small sample size and loss of children to follow up. While it has been suggested that average age at diagnosis in those subsequently needing operative treatment for CDH could be used at district level to monitor the outcome of screening,[246] the small numbers expected in any one district may preclude meaningful interpretation. Local audit of the screening programme is a time-consuming and laborious process[65,127] and although broad measures to monitor the current screening programme have been recommended,[1] there has been a failure to implement these,[234] reflecting a lack of agreed measures of process and outcome and a paucity of information systems to facilitate audit.[88,127] Monitoring of coverage is an important issue, especially for a community-based programme and yield at each examination should be recorded.

Whatever the shape of the future programme, measures of process and outcome are needed on a national basis. The current study showed that a national estimate of the prevalence of a first operative procedure from an active reporting scheme alone would be unreliable, and time consuming to compile and validate. Furthermore, active reporting on an ongoing basis may not be feasible in view of the high clinical workload of orthopaedic surgeons. HES data may provide an alternative and adequate

means of monitoring the prevalence of surgery. However, inspection of a sample or all of the medical records would be required to identify the proportion of children not detected by the screening programme and the proportion failing early treatment. In the current study only 48% of HES-identified children met the study case definition, although examination of the Scottish data alone suggested that this could be improved from 53% to 66% without loss of sensitivity. Further work is required to estimate the sensitivity and specificity and thus the potential of HES data to provide routine national monitoring of the CDH screening programme on a national basis, using the prevalence of surgery as a proxy outcome measure.

6.4 Implications for clinical practice

The poor sensitivity of the O-B test means that a negative result is not wholly reassuring and that vigilant surveillance of children's hips should be continued. Parental concerns led to the identification of 37% of the children who received a first operative procedure for CDH in the current study and parents should be encouraged to express their concerns. Although there is no clear evidence regarding the optimum time of treatment, expert clinical opinion suggests that prognosis is better for children detected before weight-bearing age and therefore efforts should be directed towards detecting CDH before a child's first birthday. This question will be addressed formally in a planned follow up study of children identified through the validation exercise undertaken as part of the current study.

6.5 Implications for future research

The current study demonstrated that a surveillance scheme can be set up quickly among clinicians without previous experience of a reporting scheme. However, the

rate of return of reporting cards does not necessarily reflect the completeness of case reporting. The value of multiple data sources and capture-recapture analysis was shown in this study, as has been demonstrated in similar epidemiological studies. The lack of a second source of cases of abduction splinting is consequently a concern. This might be addressed by a more intensive surveillance in a smaller number of maternity units. A prospective reporting scheme in which identifying details were requested at the time of reporting may have been more suitable for the identification of cases of abduction splinting.

Knowledge of the reported screening and management practices and nationally representative data on the prevalence of treatment for CDH provided important data from which to judge the public health importance of CDH and the overall effectiveness of the screening programme. This work has provided up to date, and representative data from which to develop future trials and related research. It provides some of the key parameters for a decision analysis which will inform potential future trials of screening methods. Outcome in relation to age at diagnosis, at first operative procedure and complexity of first operative procedure will be addressed in a representative sample of the current study population. Examination of the inter- and intra-observer variation in the reporting of ultrasound images will be examined in a further study.

This study highlights the difficulties in evaluating an established screening programme for which direct evidence of effectiveness is lacking. This situation is common to the screening for conditions other than CDH, for example, vision screening. Observational epidemiological data can provide an important contribution to evaluation, particularly

for questions which experimental methods have not addressed, or to which they may not easily be applied.

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ACKNOWLEDGEMENTS

I am very grateful to the paediatricians, orthopaedic surgeons, radiologists, nurses, physiotherapists and administrative staff who located notes and completed forms for this study. In addition, I am grateful to the following:

Myer Glickman for his advice during the establishment of the original OS scheme reporting base; Richard Lynn for the BPA manpower census data and his enthusiasm and support throughout the study; Anne Meade, Carol Knowles and David Adams of the British Orthopaedic Association for the BOA membership list and BOA manpower census data; Jack Vize of ISD, for his patience with my requests for successive lists of Scottish episode data; Henry Nguyen of Wessex Regional Health Authority for providing data two days before his post was abolished; Christine Ireland for completing validation forms for Wessex; Sue Fritz for the miles she travelled in completing validation forms for the Northern region and Scotland, and her commitment and accuracy; the MRC Working Party, particularly Edmund Hey for assistance in obtaining data from the Northern Regional Health Authority, Nicholas Clarke for the coding frame for complexity of surgery, and Sheila Gore for her suggestions regarding the capture recapture analysis. Professor Catherine Peckham has supported and encouraged me throughout my time at the Institute of Child Health and, she and Jugnoo Rahi, provided constructive comments on an earlier draft of this thesis. Additional criticism of my grammar and punctuation was contributed by Ben Linstead.

I am most grateful to my academic supervisor, Dr Carol Dezateux, who developed the original study design, in collaboration with the MRC Working Party, and appointed me to the study. Carol encouraged me to develop my own ideas and allowed me time to

explore, learn and progress. Without Carol's generosity, patience, and commitment to my career, this thesis could not have been written.

Thanks are also due to my flat-mate Caroline Coombes and to my family, who gave of their time to allow me time to write.

Appendix 2.1 current and past membership of the Medical Research Council Working Party

Eva Alberman (former chairperson)
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*Sue Banton
*George Bennet
*Nicholas Clarke
Christina Davies
*Carol Dezateux
*Diana Elbourne
*Frances Gardner
*Alastair Gray
*Sheila Gore
*Adrian Grant
*David Hall
Susan Hall
*Edmund Hey
Charles Kaye (former chairperson)
Leslie Klenerman
George Knox
Ian Leck
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Anne Ludbrooke
*Nicholas Mays (current chairperson)
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Carthage Smith (MRC)
Madge Vickers (MRC)

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Appendix 3.1 questionnaire, survey of screening and management practices

PAGE 1

SURVEY OF SCREENING FOR CONGENITAL DISLOCATION OF THE HIP

MRC National Study of Congenital Dislocation of the Hip
Institute of Child Health, London

This confidential questionnaire concerns screening for and management of congenital dislocation of the hip (CDH) in infants for whom you are a consultant responsible for neonatal care.
Only aggregate data will be presented and no individual clinician or hospital will be identifiable in the published report.

Please tick boxes as appropriate.

1. In case there are any queries about the replies to this questionnaire please give details of person completing form:

   Name: ☐ Telephone number: ☐

   (BLOCK CAPITALS PLEASE)

   1a. Have you been a consultant responsible for neonatal care during the last year? Yes ☐ No ☐

   If no, please return questionnaire using address given on page 7

2. Please give the name(s) and postcode of all maternity unit(s), including GP units, for which you are the consultant paediatrician responsible for neonatal care:

   (if known)

   Hospital 1: ..... Postcode ☐
   Hospital 2: ..... Postcode ☐
   Hospital 3: ..... Postcode ☐

   2a. Please give the **approximate** number of live births in the above hospital(s) for 1992:

   Hospital 1: ☐ live births Hospital 2: ☐ live births Hospital 3: ☐ live births

3. Does a person within your health district have overall responsibility to keep the hip screening programme under review? Yes ☐ No ☐ Don't know ☐

   If no or not known, please go to page 2 and complete rest of form for infants born at Hospital 1 only.

   3a. In case further information is required give name, job title and contact address of this person:

   Name ..... Job title .....

   Contact address .....

   .....

IMPORTANT: The rest of this form applies to infants born at Hospital 1 only

Appendix 3.1 questionnaire, survey of screening and management practices /c

PAGE 2

Questions 4 to 6 concern screening for congenital dislocation of the hip by clinical examination in term infants born in Hospital 1.

4. When is the first routine clinical examination of the hips after birth usually performed?

In the first 24 hours of life ☐ In the first 48 hrs of life ☐ After 48 hours of life ☐: please specify age ☐ days

4a. Who usually performs the first clinical examination of the hips after birth?
(You may tick more than one box)

Paediatric SHO ☐ Paediatric Reg ☐ Obstetric SHO ☐ Obstetric Reg ☐
Midwife ☐ General Practitioner ☐ Physiotherapist ☐ Clinical Medical Officer ☐

other ☐ *(please give job title)* .....
don't know ☐

5. Is a second routine clinical examination of the hips ever performed in term infants before discharge? Yes ☐ No ☐ Don't know ☐
If no or not known, go to question 6

5a. In approximately what percentage of term infants born at Hospital 1 is this second routine examination performed? ☐ % Don't know ☐

5b. What determines which infants receive a second routine examination before discharge?
(You may tick more than one box)

Above certain age at discharge ☐ : please specify this age ☐ hours

other ☐ *(describe)* .....
don't know ☐

5c. Who usually performs this second routine examination?
(You may tick more than one box)

Paediatric SHO ☐ Paediatric Reg ☐ Obstetric SHO ☐ Obstetric Reg ☐
Midwife ☐ General Practitioner ☐ Physiotherapist ☐ Clinical Medical Officer ☐

other ☐ *(please give job title)* .....
don't know ☐

6. What training in hip examination is usually offered to the staff who perform the first routine clinical hip examination, when they start work in Hospital 1? *(You may tick more than one box)*

clinical/bedside teaching ☐ by? *(job title)* .....
training with Baby Hippy ☐ how frequently?: always ☐ sometimes ☐ rarely ☐
other ☐ *(describe)* .....
.....
none ☐
don't know ☐

Appendix 3.1 questionnaire, survey of screening and management practices /c

PAGE 3

Questions 7 to 10 concern ultrasound examination of the hips of infants born in Hospital 1.

7. Do any term infants born in Hospital 1 undergo ultrasound examination of the hips for screening or diagnostic purposes? Yes ☐ No ☐
If yes, please go to Q.8

7a. Is there any plan to introduce ultrasound examination of the hips for screening or diagnostic purposes in the next 12 months? Yes ☐ No ☐ Don't know ☐
If ultrasound not used, go to page 4

8. In which year was ultrasound examination of the hips introduced at Hospital 1? 1 9 ☐ ☐ *(enter year)* Don't know ☐

8a. How is ultrasound examination of the hips used in Hospital 1? *(You may tick more than one box)*

to screen all term infants for CDH ☐
to screen infants considered to be at high risk of CDH ☐
to further assess infants with clinically detected hip abnormalities ☐
other ☐ *(please describe)* .....

8b. If not currently used for screening purposes, is there any plan to introduce hip screening by ultrasound at Hospital 1 in the next 12 months? Yes ☐ No ☐ Don't know ☐ Not applicable ☐

9. Who usually performs the ultrasound examination? *(You may tick more than one box)*

Consultant Radiologist ☐ Consultant Orthopaedic Surgeon ☐ Radiographer ☐
Radiology SR ☐ Orthopaedic SR ☐ Don't know ☐
other ☐ *(please give job title)* .....

9a. Who usually reports the ultrasound? *(You may tick more than one box)*

Consultant Radiologist ☐ Consultant Orthopaedic Surgeon ☐
Radiology SR ☐ Orthopaedic SR ☐ Don't know ☐
other ☐ *(please give job title)* .....
not formally reported ☐ *(please describe what happens)* .....
.....

9b. What type of ultrasound examination is usually performed? static ☐ dynamic ☐ not known ☐
(You may tick more than one box)

9c. How is the static ultrasound examination usually reported? *(Tick one box only)*

not applicable ☐ by measuring angles and using Graf's classification ☐
don't know ☐ by measuring angles and using a modified Graf classification ☐
other ☐ *(please describe)* .....

10. Please give name, job title and contact address of person primarily responsible for the hip ultrasound service, in case further information is required:

Name ..... Job title .....

Contact address .....

.....

Appendix 3.1 questionnaire, survey of screening and management practices /c

PAGE 4

Questions 11 to 14 concern the screening and management of infants born in Hospital 1 considered to be at high risk of congenital dislocation of the hip.

11. Are infants born in Hospital 1 and considered to be at high risk of congenital dislocation of the hip screened or managed differently to those infants not at high risk? Yes ☐ No ☐ Don't know ☐
If no or not known, go to page 5

11a. Which of the following criteria are used to identify a high risk group who are screened or managed differently?

(Please tick Yes, No or Don't know for each item)

	Yes	No	Don't know		Yes	No
family history of dislocated hips	☐	☐	☐	If yes, is this for first degree relatives only	☐	☐
family history of "clicking" hips	☐	☐	☐	If yes, is this for first degree relatives only	☐	☐
oligohydramnios	☐	☐	☐			
breech presentation	☐	☐	☐			
caesarean section	☐	☐	☐			
intrauterine growth retardation	☐	☐	☐			
talipes	☐	☐	☐			
other congenital abnormality	☐	☐	☐	If yes, which		
other condition	☐	☐	☐	If yes, which		

12. Which of the following are usually done, and at what approximate age, to further assess this high risk group?
*(tick all boxes which apply, *delete as applicable and insert age where shown)*

further clinical assessment by paediatric consultant/SR*	☐	at ☐	days/weeks/months of age*
further clinical assessment by orthopaedic consultant/SR*	☐	at ☐	days/weeks/months of age*
ultrasound examination of the hip	☐	at ☐	days/weeks/months of age*
radiological examination of the hip	☐	at ☐	days/weeks/months of age*
other *(describe)*.....	☐	at ☐	days/weeks/months of age*
don't know	☐		

13. Are the assessment(s) identified in Question 12 performed in all high risk infants born at Hospital 1? Yes ☐ No ☐ Don't know ☐

13a. If no, please describe the criteria used to select which infants are assessed? Not applicable ☐

.....

.....

.....

14. Are these assessments performed even if a high risk infant has a normal first or second routine clinical hip examination? Yes ☐ No ☐ Don't know ☐

Appendix 3.1 questionnaire, survey of screening and management practices /c

Questions 15 to 20 concern the management of infants born at Hospital 1 in whom a dislocated or dislocatable hip is suspected before discharge.

15. a. If a dislocated hip is suspected before discharge, which of the following are performed before deciding to treat?
*(tick all which apply, *delete as applicable and insert age where shown)*

clinical re-examination by paediatric consultant/SR/Reg* ☐ at ☐ days/weeks of age*
clinical re-examination by orthopaedic consultant/SR/Reg* ☐ at ☐ days/weeks of age*
ultrasound examination ☐ treated immediately ☐ don't know ☐

other ☐ *(please describe)* .....

.....

b. If a dislocatable hip is suspected before discharge, which of the following are performed before deciding to treat?
*(tick all which apply, *delete as applicable and insert age where shown)*

clinical re-examination by paediatric consultant/SR/Reg* ☐ at ☐ days/weeks of age*
clinical re-examination by orthopaedic consultant/SR/Reg* ☐ at ☐ days/weeks of age*
ultrasound examination ☐ treated immediately ☐ don't know ☐

other ☐ *(please describe)* .....

.....

16. a. Once the diagnosis of a dislocated hip is confirmed, who usually decides to start treatment?

orthopaedic surgeon ☐ *(give grade)* .....
paediatrician ☐ *(give grade)* .....
other ☐ *(give job title)* .....
don't know ☐

b. Once the diagnosis of a dislocatable hip is confirmed, who usually decides to start treatment?

orthopaedic surgeon ☐ *(give grade)* .....
paediatrician ☐ *(give grade)* .....
other ☐ *(give job title)* .....
don't know ☐

17. a. Which of the following are usually used, and for approximately how long, as initial treatment for a confirmed dislocated hip?

splint appliance ☐ *(give types)* .....
..... for ☐ weeks
plaster of Paris ☐ for ☐ weeks
other ☐ *(describe)* .....
..... for ☐ weeks
don't know ☐

Appendix 3.1 questionnaire, survey of screening and management practices /c

PAGE 6

17. b. Which of the following are usually used, and for approximately how long, as initial treatment for a confirmed dislocatable hip?

splint appliance ☐ *(give types)*..... for ☐ weeks

.....

plaster of Paris ☐ for ☐ weeks

other ☐ *(describe)*..... for ☐ weeks

.....

don't know ☐

18. Please give full job title (including specialty) of person who usually fits the splint appliance:

Not applicable ☐ Don't know ☐

Full job title: .....

19. a. Is ultrasound used to monitor progress once treated? Yes ☐ No ☐ Don't know ☐

b. Is ultrasound examination used to determine duration of treatment? Yes ☐ No ☐ Don't know ☐

20. Is further assessment arranged for infants with a dislocatable or dislocated hip, that has stabilised spontaneously without treatment?

Yes ☐ No ☐ Don't know ☐

If yes, please describe assessment .....

.....

21. a. Are double or triple nappies ever used to manage infants with suspected hip abnormalities in Hospital 1?

Yes ☐ No ☐ Don't know ☐

b. If yes, what are the indications for using double or triple nappies?

established dislocation ☐

unstable hips ☐ "clicky" hips ☐

as interim treatment until diagnosis can be confirmed ☐

other *(please describe)*..... ☐

22. In case further information is required, please give name, specialty and contact address of person primarily responsible for the management of infants with dislocated or dislocatable hips:

Name ..... Specialty .....

Contact address .....

.....

23. Are the screening and management practices you have described on this form for Hospital 1 also applicable to:

Hospital 2? Yes ☐ No ☐ Don't know ☐ Not applicable ☐

Hospital 3? Yes ☐ No ☐ Don't know ☐ Not applicable ☐

Appendix 3.1 questionnaire, survey of screening and management practices /c

Thank you very much for taking the time to complete this form.

Please use the space at the bottom of the page to make any further comments.
Then please return this form, using the enclosed pre-paid envelope, to:

The MRC National Study of Congenital Dislocation of the Hip
Institute of Child Health
Freepost WC5645
London
WC1N 1BR

If you have any queries, please contact:

Dr Carol Dezateux or Ms Sara Godward
Telephone: 071-242 9879 ext.2605
Fax: 071-831 0488

Further comments:

Appendix 3.2 covering letter, survey of screening and management practices

Dr XX
Consultant Neonatologist
Special Care Baby Unit
XX Hospital
Town
County
Postcode

our ref: 001XX

10th December 1993

Dear Dr XX

I am writing to you in your capacity as a consultant paediatrician responsible for neonatal cover as listed in the Directory of Emergency and Special Care Units. I would be grateful for your help in this national survey of congenital dislocation of the hip (CDH), which aims to establish the relative popularity of different screening and management practices for CDH throughout the UK and the Irish Republic. This survey forms part of a review of screening and management for CDH being conducted under the auspices of the MRC Working Party.

Please would you complete the enclosed form in respect of the maternity hospitals/units for which you provide neonatal cover and return the form to me using the enclosed FREEPOST envelope. If another consultant paediatrician in your unit would be better placed to complete the form, then please pass on this letter, and the enclosed form and envelope.

A report of this survey will be published and all participants will be sent a summary of the main findings when these are available.

I am very grateful for your help in this study and look forward to receiving your form.

Yours sincerely

Dr Carol Dezateux
Senior Lecturer

British Paediatric Surveillance Unit
5 St Andrew's Place Regents Park London NW1 4LB
Tel: 071 985 1866 Fax: 071 486 6009

CONGENITAL DISLOCATION OF THE HIP

Abstract As part of a Department of Health review of population screening programmes, the MRC was asked to set up a working party in 1989 to examine existing screening policies for Congenital Dislocation of the Hip (CDH) and, in particular, the role of ultrasound imaging. This epidemiological study was considered an essential first step and will provide nationally representative data on:

1. the number of infants (per 1000 live births) receiving treatment for CDH as a result of a positive screening test;

2. the number of infants and young children (per 1000 total population) undergoing an operative procedure for CDH, in whom CDH had not been detected by screening;

3. the range and variability of existing screening practices for CDH in the UK.

This will yield important information about the screening programme as currently practised, and allow the objectives and feasibility of a trial of screening methods, including ultrasound, to be determined.

Principal Investigators Dr Carol Dezateux Senior Lecturer; Sara Godward Research Fellow

Unit of Epidemiology and Biostatistics
Division of Public Health
Institute of Child Health
30 Guilford Street
London WC1N 1EH
Telephone: 071-242 9789 ext 2605
Fax: 071-831 0488

Duration of Study April - June 1993

Geographic Area UK and Republic of Ireland

Background Congenital dislocation of the hip is one of the most common and potentially disabling congenital deformations. The current UK screening programme, introduced without prior evaluation, aims to identify pre-symptomatic cases by clinical examination when abduction splinting may avoid the need for surgery. However, the effectiveness of this programme has been questioned due to the continued presentation of cases not detected by screening. Furthermore, numbers of infants treated as a result of a positive screening test are recognised to exceed the number that would be treated for established dislocation in the absence of screening, suggesting a potentially high false positive rate. Screening by ultrasound examination has been proposed as a more effective alternative screening method, but this requires formal evaluation.

The purpose of this survey is to determine prospectively, by means of an active reporting scheme involving both paediatricians and orthopaedic surgeons, the number of infants treated for CDH as a result of a positive screening test. The number of infants and children requiring surgical treatment for CDH, who had not been detected by screening, will be ascertained by a special surveillance scheme involving orthopaedic surgeons only. This will be followed by a postal questionnaire to paediatricians responsible for maternity units to identify the range of screening practices currently employed.

Research Questions

1. How many infants are being treated for CDH as a result of a positive screening test, and how does this vary by country and Region?

2. How many infants and children are receiving operative treatment for CDH, who had not been detected by screening? How does this vary by country and Region?

3. What is the range and variability of screening practices employed in maternity units nationally and in what proportion is ultrasound examination already part of routine practice?

Case Definition Congenital Dislocation of the Hip is defined as 'a deformation, of the hip joint present at birth, in which the head of the femur is, or may be, partly or completely displaced from the acetabulum. This includes secondary hip joint dysplasia whether or not hip instability or dislocation persists' (Dunn).

Reporting Instructions Paediatricians are requested to report all infants known to them in whom abduction splinting (including double nappies) for CDH was initiated during the preceding month.

Ethical Approval Ethical approval has been obtained for this study from the Ethical Committee of the Hospital for Sick Children, Great Ormond Street.

Workload It is estimated that approximately 2,000 cases will be notified during a 3 month period of surveillance for an average treatment rate of 10 per 1,000 live births. This represents an average reporting burden of two per paediatrician over a three month period. The reporting paediatrician will be asked to complete a short questionnaire for each new case.

Funding The study is funded by the Medical Research Council.

Reference List available from the BPSU office on request.

Administrator: Mr Richard Lynn, MSc
British Paediatric Surveillance Unit
5 St Andrew's Place Regents Park London NW1 4LB
Tel: 071 985 1866 Fax: 071 486 6009

CONGENITAL DISLOCATION OF THE HIP

Background

Congenital dislocation of the hip is one of the most common and potentially disabling congenital deformations. The current UK screening programme, introduced without prior evaluation, aims to identify pre-symptomatic cases by clinical examination when abduction splinting may avoid the need for surgery. However, the effectiveness of this programme has been questioned due to the continued presentation of cases not detected by screening. Furthermore, numbers of infants treated as a result of a positive screening test are recognised to exceed the number that would be treated for established dislocation in the absence of screening, suggesting a potentially high false positive rate. Screening by ultrasound examination has been proposed as a more effective alternative screening method, but this requires formal evaluation.

In view of this, and at the request of the Department of Health, the MRC established a working party to examine existing screening policies for Congenital Dislocation of the Hip (CDH) and, in particular, the role of ultrasound imaging. This epidemiological study was considered an essential first step. It aims to provide nationally representative data, covering the UK and Eire, to answer the following research questions:

Research Questions

1. How many infants per 1,000 live births are being treated for CDH as a result of a positive screening test, and how does this vary by country and region?

2. How many infants and children aged 5 and under are receiving operative treatment for CDH? What proportion of these had not been previously detected by screening? How do these figures vary by country and region?

3. What is the range and variability of screening practices employed in maternity units, and in what proportion of centres is ultrasound examination already part of routine practice?

How will this information be obtained?

The numbers of infants and young children with CDH requiring treatment with abduction splinting, double nappies or a first operative procedure will be determined prospectively by means of an active reporting scheme involving paediatricians and orthopaedic surgeons. A scheme for paediatricians - the British Paediatric Surveillance Unit (BPSU) - is already in place and has been successfully used to obtain population-based data on a variety of paediatric conditions. With the collaboration of the British Orthopaedic Association (BOA), a similar active reporting scheme for orthopaedic surgeons has been established specifically for this study. This scheme will be administered by the BPSU (Administrator, Mr Richard Lynn). Consultant orthopaedic surgeons in the UK and Eire who treat children for CDH, albeit rarely, have been identified by a prior postal survey.

Orthopaedic surgeons thus identified will participate in the reporting scheme as described below.

What is required of participating surgeons?

Orthopaedic surgeons will be asked to report cases of CDH to the BOA surveillance scheme as follows:

At the end of each month of the study, each surgeon will receive, from the BPSU, a green card consisting of two parts separated by a perforated strip. They will be asked to post the reply-paid portion of this card to the BPSU indicating in the boxes where appropriate:

- the number of infants with CDH in whom treatment with abduction splinting or double nappies was started in that month;
- the number of infants and young children with CDH aged 5 and under receiving a first operative procedure for CDH in that month, with or without general anaesthesia.

The card should be returned even if no cases have been seen, and this may be indicated by ticking the "Nothing to report" box.

The second half of the card is retained by the consultant and may be used to record information to assist identification of reported cases. Consultants reporting a case will be contacted subsequently by the study investigators and asked to complete a one-page questionnaire for each case reported.

Study timetable and duration

Cases of treatment with abduction splinting and double nappies will be ascertained through paediatricians and orthopaedic surgeons participating in the BPSU and BOA schemes respectively for a 3 month period from April to June 1993 inclusive.

Cases of a first operative procedure for CDH will be ascertained through the BOA scheme for 12 months from April 1993 to March 1994.

Information on screening and management for CDH will be obtained by a postal survey in late 1993.

Study investigators

Dr Carol Dezateux — Senior Lecturer
Sara Godward — Research Fellow

Unit of Epidemiology and Biostatistics
Division of Public Health
Institute of Child Health
30 Guilford Street
London WC1N 1EH
Telephone: 071-242 9789 ext 2605 Fax: 071-831 0488

Ethical Approval

Ethical approval has been obtained for this study from the Ethical Committee of the Hospital for Sick Children, Great Ormond Street.

Funding

The study is funded by the Medical Research Council.

References

List available from BPSU on request.

Appendix 3.5 article published in the *British Orthopaedic News*, spring 1993

MRC National Study of Screening and Treatment for Congenital Dislocation of the Hip

A Working Party on Congenital Dislocation of the Hip (CDH) has been convened at the request of the Department of Health to review the current national screening programme for CDH. This reflects concern about the continued presentation of late cases of CDH as well as increasing interest in the use of ultrasound examination of the hips as a screening test.

A national survey of screening and treatment for CDH was considered an essential first step. The purpose of this study is to establish the range and variation in current screening practices in the UK and Ireland and to identify the number of infants and young children requiring treatment for CDH.

This study is being carried out in collaboration with the British Orthopaedic Association (BOA) and the British Paediatric Surveillance Unit (BPSU). The support of both orthopaedic surgeons and paediatricians is crucial to its success.

Fellows of the British Orthopaedic Association will be contacted this month and asked whether they treat children, however rarely. Members thus identified will be contacted in April with further details of the study. They will be asked to take part in an active reporting scheme similar to that successfully run by the BPSU. Participating members will be asked to indicate the *number of cases of CDH in whom treatment was started in the preceding month* by ticking the relevant box on a reply-paid postcard. A new card will be sent each month for a 12-month period starting in April 1993. The card includes a "nothing to report" box and *it is vital that each member return the card each month even if there is nothing to report.* Members reporting a case will be asked to complete a *brief* questionnaire giving more details of the case.

Further information will be forthcoming in the BOA mailing and a progress report is planned for the Autumn issue of *British Orthopaedic News*.

More information may also be obtained from Dr Carol Dezateux who is co-ordinating this project, or from the orthopaedic members of the MRC Working Party: Professor Leslie Klenerman and Mr Nicholas Clarke (addresses given below).

Contacts:
Dr Carol Dezateux, MRCP
Senior Lecturer and Research Co-ordinator
Unit of Epidemiology
Institute of Child Health
30 Guilford Street
London WC1N 1EH
Tel: 071-242 9789, ext 2605

Professor Leslie Klenerman, FRCS
University Department of Orthopaedic and Accident Surgery
Royal Liverpool University Hospital
Prescott Street
PO Box 147
Liverpool, L69 3BX
Tel: 051-706 4122

Mr Nicholas Clarke, FRCS
Southampton General Hospital
Tremona Road
Southampton, SO9 4XY
Tel: 0703 777222

National Tuberculosis Survey*

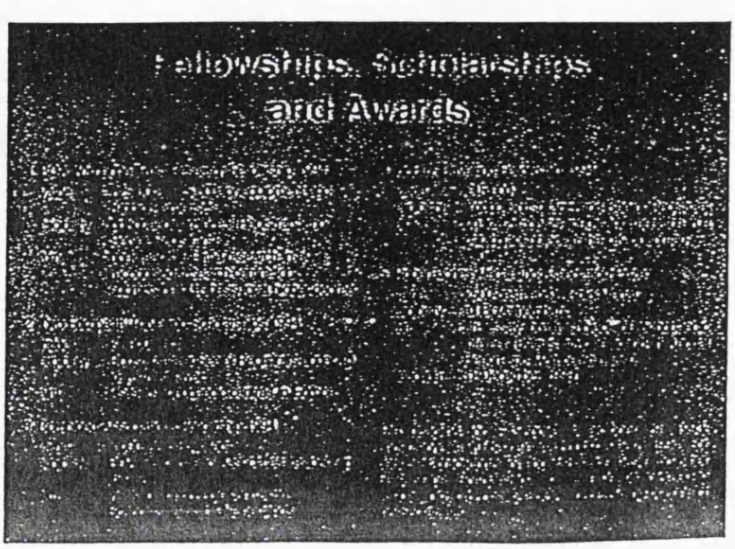
A National Tuberculosis Notifications Survey is planned for 1993 for England and Wales, co-ordinated from PHLS Communicable Disease Surveillance Centre in collaboration with the British Thoracic Society and the Department of Health. Three similar surveys have been undertaken by the Medical Research Council in the past and have provided information on recent trends in tuberculosis not available from routine surveillance.

The 1993 survey will estimate the occurrence of tuberculosis in a range of population sub-groups and the trends in these groups over time, and will collect information on clinical features and treatment. This information is essential for decisions about future control measures, including the schools' BCG programme. A special aspect of the survey on this occasion is the estimation of HIV prevalence amongst adults (16-54 years) notified with tuberculosis, using the unlinked anonymous HIV testing methodology which is now well established.

During the survey period (December 14, 1992 to December 31, 1993) all cases of tuberculosis notified to the 'proper officer' (usually the Consultant in Communicable Disease Control [CCDC]) will be included. During the last six months of 1993, the survey will be restricted to cases under 55 years of age only. We would therefore like to remind Fellows about the importance of notifying all newly diagnosed cases of bone and joint tuberculosis and to ask for your co-operation in completing the survey 'Clinical Form' that will be sent to you. We would also seek your co-operation in including eligible patients in the HIV Prevalence Survey. All survey data will be held in strict medical confidence.

Further details, including instructions for including patients in the HIV Prevalence Component, will be sent to clinicians who notify cases of tuberculosis during the study period.

*National Survey of Notifications of Tuberculosis in England and Wales in 1993 — a Public Health Laboratory Service/British Thoracic Society/Department of Health collaborative study.



BOA REPORT CARD

FEBRUARY 1994 [93]

CODE No :

☐ Please tick if **NO CASES TO REPORT**

☐ number of cases of a FIRST OPERATIVE PROCEDURE

Please report the number of children with CDH aged under 5 receiving a first operative procedure for CDH with or without general anaesthesia in the last month.

PLEASE RETURN THE CARD EVEN IF YOU HAVE NOTHING TO REPORT

MRC National Study of Congenital Dislocation of the Hip

BOA REPORT CARD - cases seen in February 1994

Please keep a record of the case(s) you have notified for easy reference when you are contacted by the investigator.

Patient's name &
Hospital Number

DETACH THIS SECTION BEFORE POSTING

MRC National Study of Congenital Dislocation of the Hip

BOA REPORT CARD - cases seen in February 1994

Please write the number of cases seen this month in the appropriate box. If you have not treated a child with CDH by surgery, please tick **NOTHING TO REPORT** and return the card. Thank you.

MRC National Study of
Congenital Dislocation of the Hip
Institute of Child Health
FREEPOST WC5645
LONDON
WC1N 1BR

Appendix 3.7 advance notification to paediatricians of CDH study

BPA NEWSLETTER

March 1993

Inside this issue...

You are what you eat:
Gill Cawdron describes a new project that aims to teach schoolchildren about healthy diet.
page 2

Advance Australia, Phaire:
or how the Boke's author made it good down under.
See Out and About,
page 4

The college debate:
Sir David Hull, president of the BPA, discusses the vote to seek new status as a college.
page 6

Plus:
Letters, Who's Who, Spock.
page 7

British Paediatric Association

Keeping members in touch

Welcome to the new-style *BPA Newsletter*.

Its predecessor was worthy and bulkier, but we feel that it had elements of the parish magazine.

Its replacement hopes to be informative and entertaining. We look forward to receiving news items, letters, photos or misprints. Send them to Patricia Kershaw at the BPA Office, 5 St Andrew's Place, Regents Park, London NW1 4LB.

If you would like to submit a feature article (*see page* 2) please ring me on 0295 229014.■

Harvey Marcovitch, Hon Editor

New Year's honour for Sir David Hull

BPA president, Professor David Hull, became a knight following the publication of the New Year's honours list.

Sir David commented: 'I have had the good fortune to work with a fine group of people who share my concern in developing medical services for children. I would like to think the award is a general recognition of what we are trying to do.'

Dame June Lloyd, the past president, told the *Newsletter* that she was delighted by the news, and saw it as a just reward for hard work put in by Sir David and his team into developing both hospital and community paediatric services in Nottingham.

SIR DAVID HULL: 'good fortune'

Dr Christopher Nourse, BPA treasurer, emphasised Sir David's work in Nottingham, but added that it also represented recognition of his excellent work on the Cleveland committee of enquiry. 'His knighthood is a boost to paediatrics in general', he said.■

Hip screening project starts

Congenital hip dislocation screening will be examined as part of a national study starting in April.

The BPSU and the British Orthopaedic Association are to collaborate in data collection.

Study coordinator is Dr Carol Dezateux from the Institute of Child Health. Interest in the role of ultrasound and concern at late diagnosis led the Department of Health to set up a working party on CDH.

Dr Dezateux says that it decided the first steps would be to discover how established screening techniques are used in the UK and Ireland and how many infants are picked up or missed.

As with all BPSU projects, the cooperation of BPA members is essential. A member of the study team will be available at the Warwick meeting.■

Appendix 3.8 questionnaire sent to surgeons identified by manpower census

Mr XX
Consultant orthopaedic surgeon
XX Hospital
XX Road Street
Town
County
Postcode

31st March 1995

Dear Mr XX

In collaboration with the British Orthopaedic Association (BOA), reports of cases of congenital dislocation of the hip (CDH) in the months April 1993 to April 1994 (incl) were obtained from orthopaedic surgeons identified as Home Fellows of the BOA. This was to provide nationally representative figures for the incidence of CDH, the first step of the programme of the MRC Working Party on CDH.

Your name was subsequently identified from the BOA manpower census data but was not apparently included in the BOA Home Fellows list. I should therefore be grateful if you would indicate on the form below whether you were practising as a consultant orthopaedic surgeon during the period of the study and if so, whether this involved the treatment of children with CDH. This will allow us to estimate the completeness of the reporting base. Please return this letter using the reply-paid envelope enclosed.

Thanking you in anticipation.
Yours sincerely

Dr Carol Dezateux
Senior Lecturer

Please tick the appropriate boxes Mr XX

Did you practise as a **consultant** orthopaedic surgeon during **all or part** of the period April 1993 to April 1994?

Yes ☐ No ☐ *If no*, please return this letter now

If yes, Was this at the above hospital?
Yes ☐ No ☐ *If no,* please name hospital and town:

Did you treat children for CDH with either splint appliances or surgery (including arthrograms, PoP casts) in the period April 1993 to April 1994?
Yes ☐ No ☐

Thank you for completing this form. Please return it to Please return to: MRC National Study of CDH, Institute of Child Health, FREEPOST WC5645, London, WC1N 1BR

Appendix 3.9 questionnaire sent to paediatricians identified by manpower census

Dr XX
Consultant Paediatrician
XX Hospital
XX Road
Town
County
Postcode

3rd May 1995

Dear Dr XX

In collaboration with the British Paediatric Surveillance Unit (BPSU), cases of congenital dislocation of the hip (CDH) were reported by consultant paedatricians included in this scheme during the period April to July 1993 (incl.). This was to obtain nationally representative figures for the incidence of CDH, the first step of the programme of the MRC Working Party on CDH.

Your name was subsequently identified from the BPA manpower census data but was not apparently included in the BPSU list. I should therefore be grateful if you would indicate on the form below whether you were practising as a consultant paediatrician during the period of the study and if so, whether this involved the treatment of children with CDH. This will allow us to estimate the completeness of the reporting base. Please return this letter using the reply-paid envelope enclosed.

Thanking you in anticipation.
Yours sincerely

Dr Carol Dezateux
Senior Lecturer

Please tick the appropriate boxes Dr XX

Did you practise as a **consultant** paediatrician during **all or part** of the period April to July 1993?

Yes □ No □ *If no,* please return this letter now

If yes, Was this at the above hospital?

Yes □ No □ *If no,* please name hospital and town:

Please state any area of special interest while in this post, eg neonatology.....

Did you treat children for CDH with splint appliances in the period April to July 1993?

Yes □ No □

Thank you for completing this form. Please return it to: MRC National Study of CDH, Institute of Child Health FREEPOST WC5645, London, WC1N 1BR

Appendix 3.10 follow up form for notified cases of abduction splinting

Follow-up questionnaire for reported cases of **abduction splinting**
MRC National Study of Congenital Dislocation of the Hip

Study number (for office use only)

Please tick boxes, complete, (ring) or delete as appropriate.
If not known, write n/k

Name of person completing form ____ Tel. No ____

Infant's first name
Infant's surname
Hospital of birth:
Name
Town
Postcode
Infant details: d d m m y y
Date of Birth Sex M / F
Ethnic group *Black/Asian/Oriental/White/Other/Not known*
Birthweight kg Gestation weeks

Mother's first name
Mother's surname
Mother's maiden name
Mother's hospital number
GP Name
GP Town
GP Postcode
Consultant responsible for continuing care:
Consultant's name
Infant's hospital number
Hospital name
Hospital town
Hospital postcode

Specify affected hip(s): *left/right/both/not known*
Age at which CDH first suspected
Were double nappies used? *yes/no* *If yes*, were they used *before/without* formal splintage? *(please ring)*
Which type of formal splint was first used?
Date splinting started d d m m y y

Was CDH detected by screening? *yes/no/not known*

If yes. By whom? (Job title, specialty)	*If no.* By whom? (Job title, specialty)
age detected	age detected
Test type: *clinical/ultrasound/both/other/not known*	Detected due to: *clinical concern/parental concern/other*
Test result L:	Details:
R:	

Last clinical examination **before starting treatment:**

	d d m m y y	*By whom?* (job title, specialty)	*Side:*	*Findings:*
Date:			*L/R/both*	

Were hips examined by ultrasound **before starting treatment?** *yes/no*

If yes	d d m m y y	*Type:*	*Side:*	*Findings:*
1st date:		*dynamic/static/both*	*L/R/both*	
2nd date:		*dynamic/static/both*	*L/R/both*	

Were any other tests, including Xrays, carried out **before starting treatment?** *yes/no*

If yes	d d m m y y	*Name of test:*	*Side:*	*Findings:*
Date:			*L/R/both*	
Date:			*L/R/both*	

Tick a box if the child has any of the features listed below

Family history of CDH ☐ · In *parent/sibling/not known/other:-*
Breech presentation in last trimester or at delivery ☐
Talipes ☐
First born ☐
Oligohydramnios ☐
Other congenital abnormality ☐ · Details:
Other ☐ · Details:
In none of these, please tick here ☐

IF THERE ARE ANY OTHER DETAILS WHICH YOU FEEL ARE RELEVANT, PLEASE INCLUDE THEM ON A SEPARATE SHEET
Thank you for completing this form. In case of queries, please contact Dr Carol Dezateux or Sara Godward on 071-242 9789 x2605
The bottom copy is for your records. Please return the **top copy** using the FREEPOST envelope to:
MRC National Study of CDH, Institute of Child Health, FREEPOST WC5645, LONDON WC1N 1BR.

Appendix 3.11 follow up form for notified cases of a first operative procedure

Follow-up questionnaire for reported cases of a **first operative procedure**
MRC National Study of Congenital Dislocation of the Hip

Study number (for office use on

Please tick boxes, complete, (ring) or delete as appropriate.
If not known, write n/k

Name of person completing form Tel. No

Infant's first name
Infant's surname
Infant's postcode
GP Name
GP Town
GP Postcode
Infant details: d d m m y y
Date of Birth Sex M / F
Ethnic group *Black/Asian/Oriental/White/Other/Not known*
Birthweight kg Gestation weeks

Infant's hospital number
Orthopaedic consultant
*Hospital of **treatment**:*
Name
Town
Postcode
*Hospital of **birth**:*
Name
Town
Postcode

Specify affected hip(s): *left/right/both/not known*
Date of diagnosis: d d m m y y
Name of first operative procedure
Date: d d m m y y

Was this preceded by abduction splinting? *yes/no*
If yes, type of formal splint Weeks in splint

Results of last clinical examination **before the first operative procedure.**

Date: d d m m y y	Side: *L/R/both*	Findings:

Were hips examined by ultrasound **before the first operative procedure?** *yes/no*

	Date: d d m m y y	*Type:*	*Side:*	*Findings:*
If yes 1st date:		*dynamic/static/both*	*L/R/both*	
2nd date:		*dynamic/static/both*	*L/R/both*	

Were any other tests, including Xrays, carried out **before the first operative procedure?** *yes/no*

	Date: d d m m y y	*Name of test:*	*Side:*	*Findings:*
If yes Date:			*L/R/both*	
Date:			*L/R/both*	

Tick a box if the child has any of the features listed below

Family history of CDH ▸ In *parent/sibling/not known/other:-*
Breech presentation in last trimester or at delivery
Talipes
First born
Oligohydramnios
Other congenital abnormality ▸ Details:
Other ▸ Details:
If none of these, please tick here

Was CDH detected by screening? *yes/no/not known*

If yes,	By whom? (Job title, specialty)	*If no.*	By whom? (Job title, specialty)
age detected:		age detected:	
Test type: *clinical/ultrasound/both/other/not known*		Detected due to: *clinical concern/parental concern/other*	
Test result L:		Details:	
R:			

IF THERE ARE ANY OTHER DETAILS WHICH YOU FEEL ARE RELEVANT, PLEASE INCLUDE THEM ON A SEPARATE SHEET
Thank you for completing this form. In case of queries, please contact Dr Carol Dezateux or Sara Godward on 071-242 9789 x2605
The bottom copy is for your records. Please return the **top copy** using the FREEPOST envelope to:
MRC National Study of CDH, Institute of Child Health, FREEPOST WC5645, LONDON WC1N 1BR.

Appendix 3.12 questionnaire to identify potential sources of abduction splinting

Dr XX
Institution
XX Street
Town
County
Postcode

8th September 1995

Dear Dr XX

I write to you as someone with an interest in congenital malformations. As part of a national study to estimate the prevalence of congenital dislocation of the hip (CDH), a surveillance scheme involving paediatricians and orthopaedic surgeons was established to obtain reports of babies and children who were treated for CDH between April 1993 and April 1994. I now wish to ascertain the availability of additional data sources of such children. These additional sources would allow validation of reports obtained through the active surveillance scheme.

I should be grateful if you could indicate below whether you are aware of any data source(s) of reports of infants with CDH and if so, the name and telephone number of the most appropriate person to contact for further information. I enclose a reply-paid envelope.

Thank you very much. I look forward to hearing from you.

Yours sincerely

Dr Carol Dezateux
Senior Lecturer

Dr XX

Please delete as applicable

I am aware of an additional source of data children treated for CDH. **Yes/No**

If yes, please give details:

Name of source (e.g. South West Thames Congenital Malformation Register):

Contact person:

Tel no.

Appendix 3.13 validation form

Congenital Dislocation of the Hip

Date questionnaire completed : ☐☐☐☐☐☐

MRC National Study (01.04.93 to 30.04.94)

Current Surname : _____ Previous Surname : _____

Forename (s) : _____ Date of Birth : ☐☐☐☐☐☐

Sex : ☐ Birthweight (kgs): ☐ Gestation (wks) : ☐

Birth presentation : ☐ 1 - cephalic 2 - breech 9 - not known

Birth hospital & Post code ☐ Record number : ☐

Treating hospital ☐ Record number : ☐

Please list all hip operative procedures before 30.04.94 at the treating hospital.

(An operative procedure is ANY procedure carried out in theatre whether or not an anaesthetic is required)

Date of procedure	Left hip	Right hip	Operative procedure
☐☐☐☐☐☐	☐	☐	_____
☐☐☐☐☐☐	☐	☐	_____
☐☐☐☐☐☐	☐	☐	_____
☐☐☐☐☐☐	☐	☐	_____
☐☐☐☐☐☐	☐	☐	_____
☐☐☐☐☐☐	☐	☐	_____

Please list all hip operative procedures at any other hospital before 30.04.94 Tick if none ☐

Date of procedure	Left hip	Right hip	Hospital and postcode	Consultant
☐☐☐☐☐☐	☐	☐	☐	☐

Operative procedure _____

☐☐☐☐☐☐	☐	☐	☐	☐

Operative procedure _____

Appendix 3.13 validation form /c

(NB. The first operative procedure is the first operative procedure for this child, irrespective of where or when performed.)

Was this child ever treated with traction prior to his/her first operative procedure :

☐ 1 - no 2 - yes 9 - not known

Date traction applied ☐☐ ☐☐ ☐☐

Date traction stopped ☐☐ ☐☐ ☐☐

Was this child ever fitted with a splint appliance prior to his/her first operative procedure. (NB. DO NOT include double nappies) ☐ 1 - no 2 - yes 9 - not known

What type of splint was used :

_____

_____

_____

_____

Date splint applied : ☐☐ ☐☐ ☐☐

Date splinting stopped : ☐☐ ☐☐ ☐☐

Last ultrasound examination before first operative procedure. ☐ 1 - none 9 - not known

Date : ☐☐ ☐☐ ☐☐

Result : _____

_____

Last X-ray examination before first operative procedure. ☐ 1 - none 9 - not known

Date : ☐☐ ☐☐ ☐☐

Result : _____

_____

Date of referral letter to surgeon responsible for surgery at the treating hospital : ☐☐ ☐☐ ☐☐

Referred by : ☐ 1 - GP 2 - paediatrician 3 - other surgeon 4 - other 9 - not known

If other please state : _____

Age child presented to the health service : ☐☐

How did child present : _____

First to suspect abnormality : ☐ 1 - parents 2 - G.P 3 - H.V 4 - Comm Paed. 5 - Hops. Dr 9 - not known

If Hops. Dr please state : _____

Was neonatal examination thought to be normal : ☐ 1 - no 2 - yes 9 - not known

If no describe abnormality : _____

Was 6 week examination normal : ☐ 1 - no 2 - yes 9 - not known

If no describe abnormality : _____

Is there a history of : Oligohydramnios ☐ 1 - no 2 - yes 9 - not known

Talipes : ☐ 1 - no 2 - yes 9 - not known

CDH in family (specify) ☐ 1 - no 2 - yes 9 - not known _____

Congenital abnormality or other medical problems (specify) ☐ 1 - no 2 - yes 9 - not known _____