

Lesson of the Week

Micropenis: an important early sign of congenital hypopituitarism

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Micropenis is an important sign in neonates, since it may be the only clue to the diagnosis of panhypopituitarism, a potentially lethal but eminently treatable condition.

Prenatal gonadotrophin deficiency may result in a small but normally formed penis

Case reports

Case 1—A boy infant was delivered at 37 weeks' gestation after an uncomplicated pregnancy. Birth weight was 2200 g. He was asphyxiated and successfully resuscitated but remained hypotonic. Jaundice developed on the third day of life and persisted. At 3 weeks of age hypothyroidism was diagnosed (plasma thyroxine concentration 54 nmol/l (4.2 µg/100 ml), thyroid stimulating hormone concentration 8.5 mU/l). Despite thyroxine replacement he failed to thrive and developed abdominal distension and vomiting. Laparotomy showed milk curd obstruction of the small intestine, and postoperatively he was ventilated for three days. Twelve days later he collapsed and had a hypoglycaemic convulsion. For the next three months he was fed intravenously and became hypoglycaemic when attempts were made to restart oral feeds. At the age of 4 months the diagnosis of hypopituitarism was suggested because of his micropenis. During a hypoglycaemic episode (blood glucose concentration 1.0 mmol/l; 18 mg/100 ml) the plasma cortisol value was noted to be low at 100 nmol/l (3.6 µg/100 ml) and concentrations of both plasma adrenocorticotropic hormone (93 ng/l) and growth hormone (7.0 mU/l) were inappropriately low, consistent with hypopituitarism. The hypoglycaemia resolved rapidly with hydrocortisone replacement and did not recur. Growth hormone deficiency was confirmed by a glucagon stimulation test and appreciable penile growth occurred after three injections of depot testosterone. His jaundice, which had persisted despite thyroxine replacement, resolved after initiation of the hydrocortisone.

Case 2—A boy infant (birth weight 3650 g) was delivered normally after an uncomplicated pregnancy. Shortly after birth he became hypothermic and hypoglycaemic and micropenis was noted. Three days after birth he became severely jaundiced with raised plasma transaminase activities, which persisted. Plasma thyroxine concentration was 16 nmol/l (1.2 µg/100 ml) without the appropriate thyroid stimulating hormone response (5.0 mU/l), indicating hypothyroidism of pituitary origin. Thyroid replacement was started but was inadequate. At 3 weeks of age he had several hypoglycaemic convulsions, and a normal blood glucose concentration could be maintained only

with 10% dextrose infusion. His condition deteriorated with hypothermia, hypoglycaemia, hypotension, and jaundice. A presumptive diagnosis of hypopituitarism was made and after hydrocortisone and full thyroxine replacement his blood pressure and temperature rose. Hypoglycaemia was satisfactorily controlled only when human growth hormone (2 units daily) was also given. Pretreatment 9 am plasma cortisol concentration was 50 nmol/l (1.8 µg/100 ml). Plasma growth hormone value was low (0.5 mU/l) during a hypoglycaemic episode and there was no luteinising hormone and follicle stimulating hormone response to luteinising hormone releasing hormone. These findings were consistent with multiple anterior pituitary hormone deficiencies.

Comment

The male external genitalia are formed as a result of androgens secreted by the fetal testis under the influence of placental human chorionic gonadotrophin. This process is completed by the end of the first trimester of pregnancy.¹ After the 14th week of gestation human chorionic gonadotrophin concentrations fall and pituitary luteinising hormone is necessary to maintain fetal testosterone so that growth of the penis may continue until birth.² Consequently prenatal gonadotrophin deficiency of any cause may result in a small but normally formed penis—micropenis.

Deficiencies of other anterior pituitary hormones may not be obvious at birth but symptoms, notably collapse, cyanosis, and fits, may develop during the first 24 hours. These are not specific and may be mistaken for other, more common disorders. Untreated congenital hypopituitarism, however, has a high early mortality³ and micropenis may be the only clinical indication for immediate investigation and treatment.⁴

Whereas paediatricians recognise the significance of virilisation of female genitalia as occurs in congenital adrenal hyperplasia, the importance of underdevelopment of male external genitalia may not be appreciated.⁵ In both cases reported here earlier investigation of pituitary function would have disclosed the need for hormone replacement. Persistent neonatal jaundice is a well recognised feature of thyroid deficiency, but it is not widely known that cortisol deficiency may also be responsible.⁶ In both patients the jaundice disappeared only once full replacement was given.

Standards for stretched penile length in the newborn period have been established⁷ but in true micropenis examination shows an obvious lack of erectile tissue, which may be a more valuable sign than measurement of length. Micropenis is a

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clear pointer to possible pituitary deficiency and is an important physical sign in newborn boys.

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For Debate . . .

Paramedics in the United Kingdom?

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There is no doubt that until recently the United Kingdom could boast the best ambulance service in the world. This is no longer the case. Standards have not fallen—indeed they have risen. The rest of the world, however, has caught up and in some places has overtaken us. These ambulancemen are no better trained than our own but are better equipped and allowed to do more. The obvious and best example to quote is the ambulance system now developing in the United States of America.

Twenty years ago there was no effective ambulance service in the United States. In many places any vehicle of suitable dimension was used to transport a sick or injured patient to hospital. Ironically, the most suitable vehicle to hand was usually the local hearse. It was no doubt difficult for the mortician to separate the demands of his patient and the demands of his profession. To overcome these problems many local communities developed a volunteer rescue squad. They fought fires, rescued trapped victims, and transported people to hospital. In an attempt to coordinate the many different squads, a programme of training was introduced in the early 1970s. This is now widespread and very well organised. The ambulancemen are referred to as emergency medical technicians, or EMTs. The basic emergency medical technician has between 80 and 140 hours of training, which covers basic anatomy and physiology and simple first aid. This "simple first aid" does, however, include a thorough training in cardiopulmonary resuscitation. This emphasis on control of the airway and belief in the positive effects of early cardiopulmonary resuscitation is a major contribution to the saving of life. A basic emer-

gency medical technician can then do further training to become an intermediate emergency medical technician. He has a further 60 to 200 hours of training and will be able to insert intravenous lines, obturator airways, and apply a medical antishock trouser suit. The obturator airway seals off the oesophagus and will allow air to go down only the trachea. There are many problems with this device including misplacement in the trachea and inducement of vomiting. It is rapidly losing favour in the United States. The medical antishock trouser suit is an inflatable suit put over the legs of the patient in an effort to increase peripheral resistance and thereby raise the blood pressure. It will also stabilise pelvic fractures. Having completed this training the emergency medical technician can then elect for even further training. A cardiac emergency medical technician can do all the above, but in addition can give drugs and defibrillate patients. A paramedic emergency medical technician does an extra 400 to 800 hours of training, and is then very well trained, and can give help equivalent to that of a trained physician in emergencies. The paramedics can pass endotracheal tubes, insert central venous lines, and interpret electrocardiograms to the envy of many a medical registrar.

There are problems, however, with training technicians to such a high level. As their knowledge and expertise is so wide and their capabilities are so great the medical profession feel a need to keep a close eye on their activities. Strict and lengthy rules are laid down for what they can do at the scene of an emergency, and usually any invasive action has to be preceded by permission from the local hospital. In addition, therefore, to the colossal expense of training these people and equipping them, there is the added expense of providing sophisticated radiocommunications with the hospital. Furthermore, when such a highly trained paramedic arrives at the scene, his whole training and *raison d'être* is to apply those skills that have been so hard won. Hence the main drawback with such a system is delay. When the paramedic arrives at the scene he has first of all to work through a check list. This includes taking a history and taking the patient's blood pressure, pulse, and an electro-

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