Mathematical development in Williams syndrome: A systematic review

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Abstract

Background: The current systematic review is the first to systematically explore and synthesis research to date on mathematical abilities in Williams syndrome (WS), a rare genetic disorder that results in an uneven cognitive profile. As mathematical development is complex and relies on both domain-specific and domain-general abilities, it is currently not clear what mathematical abilities have been examined in WS and also what the current gaps in this research area are. Methods and procedures: A total of 27 studies across 22 publications were identified through a systematic review search process. Results: Overall, all mathematical abilities, except for simple counting and subitizing abilities, were reported to be impaired but in line with overall mental-age abilities. However, the literature to date has not established the underlying causes of these mathematical difficulties in WS. Some studies suggested that mathematical abilities in WS follow an atypical developmental pathway with a greater reliance on verbal abilities than in typical development but coupled with impaired understanding of counting and knowledge of the number system more broadly. However, most included studies used different assessments of mathematical skills and there is a lack of studies that have examined more than one particular aspect of mathematical development within the same study. In addition, studies have often included large age ranges and small participant samples, despite the known large individual variability in WS. Conclusion: Although we know mathematical abilities in WS are impaired, this area is under-researched and there is a lack of longitudinal studies that provide insight into the cognitive mechanisms that underpin mathematical development in WS. Therefore, there is a lack of an evidence-base to inform interventions or educational practice.

Keywords: Number development; mathematics; Williams syndrome; systematic review.

Highlights

- This systematic review identified only 27 studies that have examined mathematical abilities in Williams syndrome (WS).
- The majority of the studies reported that mathematical abilities are delayed in WS and are likely to follow an atypical developmental pathway.
- There is a lack of information on the cognitive mechanisms that underpin mathematical development in WS, leading to a lack of evidence-based interventions or changes to educational practice.

What this paper adds

This is the first systematic review of studies that have examined mathematical development in Williams syndrome (WS) with aim to obtain a better understanding of the gaps in this research and as well as what conclusions can be drawn about the mathematical development in individuals with WS. Twenty-seven studies published in 22 publications were found. Although a number of these studies describe the mathematical difficulties of people with WS, especially focusing on the domain specific abilities that form the building blocks for mathematical development such as counting and large number estimation, this review shows some clear gaps in this research area, including the fact that very few studies have examined the complexity of the different cognitive mechanisms of mathematical abilities in WS and a lack of intervention studies that target mathematical abilities in this population.

Mathematical development in Williams syndrome: A systematic review

Williams syndrome (WS) is a rare neurodevelopmental disorder that occurs about 1 in 20,000 live births and is caused by a genetic deletion on the long arm of chromosome 7. Individuals with WS present with IQ scores between 42 and 68 (average IQ score = 55). More broadly, the WS cognitive profile is uneven with better language and face recognition abilities in contrast to poorer planning and visuo-spatial abilities (Martens, Wilson & Reutens, 2008).

Mathematics abilities have been repeatedly reported to be impaired in individuals with WS using a variety of different theoretical frameworks and outcome measures (Ansari, Donlan & Karmiloff-smith, 2003; Paterson, Girelli, Butterworth, & Karmiloff-Smith, 2006; O'Hearn & Landau, 2007; Udwin, Davies, & Howlin, 1996). It is broadly recognised that mathematical skills are important for attainment in school but also for employment and future life chances (Duncan et al., 2007; Watts, Duncan, Siegler, & Davis-Kean, 2014). Therefore, the difficulties in mathematics faced by individuals with WS could have far-reaching consequences.

Mathematics is a complex and multi-componential subject (Dowker, 2010) and thus mathematical development can be nuanced and potentially protracted, with not all mathematical abilities being equally impaired. In addition, in order to successfully learn and understand mathematical concepts and skills children require multiple domain-specific and domain-general skills. Therefore, dependent on the topic of interest multiple outcome measures may be used in research and multiple underlying mechanisms of learning have been investigated in previous studies. Yet, the current review is the first to examine what aspects of mathematical development have been researched in WS and what the robustness of these findings are with the aim to highlight any gaps in the research and discuss ideas for future research.

In typically developing populations, mathematical processing has been shown to rely upon a number of domain specific abilities. Studies in infants and young children have indicated that there may be two innate core domain specific systems that underpin mathematical abilities later in life (Feigenson, Dehaene & Spelke, 2004). One core system has been referred to as an object-tracking system that allows rapid estimation of small numbers of objects, referred to as subitizing. For randomly placed objects, the upper limit of perceptual estimation is 3 or 4 objects, depending on the population assessed. However, for well-rehearsed patterns, such as dice or finger digit patterns, the upper limit can be higher and this is often referred to as conceptual subitizing. Both perceptual and conceptual subitizing abilities have been argued to be fundamental skills for the development of counting, and mathematical development more broadly, in typically developing children (Desoete, 2014). In contrast, the Approximate Number System (ANS) is another core system but this allows rapid estimation of large quantities of objects, sounds, or actions. This system relies upon the ratios presented, known as Weber's fraction (w). In addition, in accordance with the distance effect, it is easier to discriminate between numbers or quantities that differ a lot, especially larger ones. Over development children become better at discriminating between smaller ratios (Halberda & Feigenson, 2008). The ANS has been argued to be important for mathematical development later in life, with some evidence that children who have more precise ANS abilities achieving higher performance on mathematical tasks, although results have been mixed (for recent reviews and meta-analysis see Chen & Li, 2014; Fazio, Bailey, Thompson, & Siegler, 2014). More advanced domain-specific skills, such as counting (Passolunghi, Vercelloni, & Schadee, 2007), adaptive and flexible strategy use (Geary &

Brown, 1991) and number familiarity (Muldoon, Towse, Simma, Perra, & Menzies, 2013) have also been identified as contributing to mathematical attainment.

In addition, domain general abilities are also required for mathematical processing. These skills include inhibition (being able to hold back a pre-potent response, Cragg & Gilmore, 2014), working memory (storing and manipulating information in mind, Passolunghi et al., 2007) and visuo-spatial abilities (making mental transformations of visual information, Uttal et al., 2013). Thus, the constellation of domain-specific and domain-general skills required for mathematical learning and processing emphasises the complexity of this subject.

Research on mathematical processing in WS has not, to date, been systematically reviewed. This means that current understanding of the supposed difficulties of this population may be skewed by two main factors. First, research with WS samples can have numerous motivating factors and can include prevalence and severity questions, testing of theoretical models of mathematical development or applied questions around intervention. The motivations of these research questions will lead research to be published in a variety of outlets, from developmental and cognitive psychology, learning disability and education journals. Therefore, an incomplete understanding of WS mathematical profile may be built due to the disparate backgrounds of the researchers (see Alcock et al., 2016 for issues in the broader field of mathematical cognition research).

Secondly, as already mentioned, mathematics is a multi-componential subject. Therefore, dependent on researchers' motivation for research questions, experience and access to resources, different outcome measures may be used across studies. In a recent systematic review, Simms, McKeaveney, Sloan and Gilmore (2019) identified that across 80 mathematical intervention studies applied to typical classrooms mainly author-generated outcome measures had been used which makes it is difficult to make any comparisons across studies. Therefore, in order to further understanding systematic searching of literature on mathematical skills in WS will enable any comparable outcome measures (if identified) to be synthesised. This is essential to develop a strong evidence base on the potential issues that individuals may face when learning mathematics.

Using a rigorous systematic review process, the current study primarily examined what types of mathematical skills have been investigated in WS. In addition, specific types of measures that have been used were identified and recorded. Secondly, the scientific robustness of these studies was examined in terms of number of participants and control groups involved. Finally, after synthesising the identified material the current review aimed to generate data driven conclusions focusing on what is understood about mathematical difficulties in individuals with WS and also highlight any gaps within the literature base for future research.

Method

Research protocol

The Preferred Reporting Items for Systematic Reviews and Meta Analyses (PRISMA) guidelines were followed for all stages of this systematic review. The protocol for the review was preregistered with Open Science Framework (https://osf.io/y7wxt) prior to the review being carried out. A protocol ensures that researchers stringently follow an outlined plan in a systematic review.

Search strategy

Using the search terms in Table 1, PsychInfo, Pubmed, Scopus, ERIC as well as ProQuest Dissertations and Thesis databases were searched for studies published between 1961 (when WS was first described) and May 2019. Google and Google Scholar were also searched using the search terms and the first 150 hits were extracted and recorded. Finally, the reference lists from all identified studies were hand searched to ensure that no potentially eligible study was missed. This search resulted in 3868 studies that were screened for eligibility after removing duplicates (See Figure 1).

Table 1: Summary of search terms

Population	("Williams syndrome")
Outcomes	(Math* OR Number OR "numerical processing" OR "numerical abilities" OR "numerical skills" OR Number skills" OR arithmetic* OR counting OR "number sense" OR magnitude OR estimation OR addition OR division OR Multiplication OR number*)

Figure 1 about here

Study selection

One reviewer (x blinded for peer review x) screened all titles and abstracts for broad relevance, and a random sample of 10% was double reviewed by a second reviewer (x blinded for peer review x). Studies were included if they: a) included a WS sample, b) explicitly reported mathematical outcomes, c) were published in English, d) narrative reviews and book chapters were only included if they reported original data. Purely qualitative studies were not included. Studies that included cross-syndrome comparisons were included but data was only extracted for the WS group and control groups. One chapter published between the selected dates referred to original data and the authors kindly sent the unpublished draft of this manuscript so that the data could be fully included in the review (Scerif, Ansari & Karmiloff-Smith, unpublished). All full-text papers were independently double reviewed (xblinded for peer reviewx and x blinded for peer reviewx).

Data extraction and synthesis

Data were extracted into a table format by one reviewer (x blinded for peer reviewx), and were checked for accuracy by a second reviewer (x blinded for peer reviewx). Extracted data included a) participant demographics, including number of participants, ages of the participants, ethnicity and information about socio-economic status (SES), type of study (cross-sectional, longitudinal or intervention), b) details about the type of control group (chronological age matched, mental age matched or other), c) the country where the study was conducted, d) details about the outcome measures and e) group means and standard deviations for the outcome measures. Data were summarised for all the studies reporting on each measurement tool, and a narrative synthesis of the methodological quality of each study was completed.

Results

Mathematical outcomes measured

In total, 27 studies published in 22 publications were found that examined a number of mathematical abilities in people with Williams syndrome. An overview of these studies is provided in Table 1.

Table 1 about here

Mathematical attainment. Of the 27 studies, seven measured overall mathematical attainment using a standardised cognitive measure, such as the **Arithmetic subtest from** the Wechsler's Intelligence Scale for Children (WISC), **Number Operations or Math Reasoning sub-test** from the Wechsler's abbreviated scales of Intelligence (WAIS), or Test of Early Mathematical Abilities (TEMA) (Howlin, Davies & Udwin, 1998; Howlin, Elison, Udwin & Stinton, 2010; O'Hearn & Landau, 2007; Steele, Brown, & Scerif, 2012; Udwin, Yule & Martin, 1987). One study measured mathematical attainment using an author-generated test (Paterson et al., 2006).

Two of these 7 studies included data from an earlier studied cohort (Udwin et al., 1987; Howlin et al., 1998) and thus, provided longitudinal insights (Udwin et al., 1996; Howlin et al., 2010). However, these studies have focused on longitudinal development of mathematical abilities in adults. Steele and colleagues (2012) included a longitudinal assessment of TEMA-iii abilities in 26 child participants with WS tested 12 months apart. This study showed that both overall numeracy abilities and cardinality skills improved over time, and cardinality skills were a unique predictor for increased mathematical abilities at the second testing time.

Overall, these studies showed that arithmetic was a particular area of difficulty in WS (Udwin et al., 1987; 1998). However, performance has been reported in other studies to be in line with non-verbal mental age (O'Hearn & Landau, 2007) or even above their overall mental age (Steele et al., 2012) and similar to other populations that have similar cognitive abilities, such as individuals with Down syndrome (Paterson et al., 2006; Steele et al., 2012). Therefore, although all studies report mathematical delays, the extent of this delay is unclear.

Non-symbolic skills (including subitising). The majority of the studies (n= 12) examined basic symbolic and non-symbolic magnitude discrimination skills as well as subitising abilities to assess whether these basic mathematical building blocks are impaired (below chronological age), spared (in line with chronological age) or atypical (not in line with abilities measured in typical populations) from infancy onwards in WS. Starting in infancy, studies showed that whilst infants with WS can discriminate between 2 and 3 objects or dots (Paterson et al., 2006; Van Herwegen, Ansari, Xu & Karmiloff-Smith, 2008), they do not show a preference for novel versus familiarised trials when shown larger sets of stimuli such as 8 versus 16 dots (Van Herwegen et al., 2008). This suggests that, whilst participants with WS are able to represent small numerosities non-symbolically, large non-symbolic representations might be impaired. Further studies in older children and adults with WS examined subitizing abilities for small set sizes (1-3) as well as larger set sizes (5-12 or 4-8) and have found that, whilst subitizing abilities for set sizes 1 to 3 are 100% accurate (albeit slower) (Ansari et al., 2007; O'Hearn et al., 2011), accuracy related to **estimation** abilities for set samples comprising larger quantities do not improve with age (Ansari et al., 2007). The subitizing abilities of adolescents and adults with WS are poorer compared to 6,5 year-old typically developing controls (O'Hearn et al., 2011). These studies suggest that the ability to present and discriminate between small set samples, being either symbolic or non-symbolic, is present from infancy onwards. However, the studies thus far have not examined the developmental trajectory of these skills, either cross-sectionally or longitudinally. Therefore, whether the development of these subitizing skills is typical remains an open question and is yet to be assessed through long-term follow-up studies.

Other studies showed that large non-symbolic magnitude discrimination abilities remain impaired into adulthood in WS and that, although there is improvement with age (Libertus, Feigenson, Halberda, & Landau, 2014), the development of approximate estimation abilities from childhood to adulthood does not extend beyond the developmental improvement seen from 4- to 7 years old (Ansari et al., 2007) and remains below the abilities seen in 6-9 year old typically developing children (Libertus et al., 2014).

One suggestion as to why people with WS might struggle with large non-symbolic estimation abilities is because of an impairment in the mental number line, or the representation of numbers in relation to space. In the number line task children are requested to indicate the position of a series of numbers on blank number lines (e.g. 0-10, 0-100; see Siegler & Opfer, 2003). Research with typically developing children consistently indicates that estimation skills are age and scale dependent, with children developing more accurate estimation patterns with age, beginning with smaller and progressing to larger scales (0-10, 0-20 then 0-100; Muldoon et al., 2013). Indeed, Paterson and colleagues (2006) have shown using a non-symbolic dot comparison task that older children and adults with WS do not display the typically observed symbolic distance effect, i.e. do not find it easier to discriminate between two sets that include a number of dots that are far removed compared to sets that are closer in number.

Symbolic skills. Opfer and Martens (2012) showed that, although adults with WS showed improvements in symbolic number line estimations compared to children with WS, their performance was still best predicted by a logarithmic and inaccurate function instead of an accurate linear one. Performance was contrasted with a much younger typically developing control group and this suggests that even adults with WS have a less optimised mental number line. However, this study included number lines from 0 to 1000 and previous studies have shown that this is beyond the number knowledge of even adults with WS. Therefore, it is difficult to conclude whether participants have impaired number line performance per se, or that the task in this study is beyond the boundary of their numerical knowledge.

Yet, further evidence of difficulties with mental number line comes from symbolic comparison tasks in which participants have to decide as quickly as possible which of two numbers are larger. Three studies have examined digit comparison in WS thus far and all used single digits (i.e. within the number knowledge range of people with WS). These studies have shown that individuals with WS respond slower in digit comparison tasks compared to second-graders and that participants with WS have a greater symbolic distance effect as well (Krajcski et al., 2009). A further study by Scerif and colleagues (unpublished) also found a difference in reaction times between the WS and a control group for close number pairs but not for distant comparisons. It is important to note that this effect was only observed for digit comparisons but not when lexical distance effect for words was examined in a rapid speeded naming tasks. This difference shows that individuals with WS experience specific difficulties with numerical representations, not with distance effects in general. However, Paterson (2000) failed to find a significant distance effect when assessing 8 children and adults with WS on a symbolic distance comparison task. The participants in this study made a larger number of errors than either of the control groups, suggesting that there is likely to be some abnormality in number processing in this group. However, these results should be treated with caution due to the extremely small sample size and large error rate for WS participants.

Counting skills. Three studies have examined counting aloud abilities in participants with WS. These studies show that participants with WS generally have good knowledge of the counting names and primary school aged children are able to count from 0-10 (Steele et al., 2012) and more broadly children and adults are able to count onwards from 1 to 20 without many problems (Ansari et al., 2007; Paterson et al., 2006). However, performance on counting tasks that require working memory abilities, such as counting onwards and backwards was poorer than typically developing controls (Paterson et al., 2006). In addition, in relation to counting sets or understanding the purpose of counting, i.e. *cardinality*, a number of studies showed that performance was delayed in line with non-verbal mental age (Ansari et al., 2003; Dolscheid & Penke, 2018; O'Hearn et al., 2011; Rousselle et al., 2013; Steele et al., 2012). This result was replicated in five studies, despite the different number of participants, ages of participants and control group(s) included. Furthermore, Ansari and colleagues (2003) showed that cardinality abilities relate to verbal abilities rather than visuo-spatial abilities as observed in a typically developing group. This would suggest that cardinality abilities in WS are not only delayed but also atypical. There is one study by Dolscheid and Penke (2018) that has also examined the understanding of quantifiers such as some, many, more etc. and has shown that, although the understanding of quantifiers is better compared to those with Down syndrome, performance was similar to younger typically developing children matched for non-verbal mental age. In addition, understanding of the word ‘some’ was extremely poor. Therefore, although individuals with WS have little problems learning the counting names and can count sets, their understanding of concepts associated with counting is delayed and atypical (Ansari et al., 2003).

Arithmetic skills. Very few studies have examined specific academic mathematical abilities such as addition, multiplication or digit recognition. Two studies have examined simple addition and multiplication. One study (Robinson & Temple, 2013) involved a case study of a teenager with WS who had good accuracy on lexical decision, naming and generating definitions for mathematical words and showed similar performance to a typically developing control group matched for verbal mental age for additional facts (despite being slower). However, performance on multiplication was significantly poorer compared to TD controls. Krajcsi and colleagues (2009) assessed a slightly larger sample of individuals with WS (n= 8) on addition and multiplication tasks and found that error rates were low and that reaction time performance in the WS group was relatively fast compared to that of 8-to-9-year old typically developing controls, despite the WS individuals being on average 17.8 years old. However, only numbers 1 to 8 were included in the calculations and none of the answers exceeded the number 16. Participants with WS have little difficulty recognising single digits, even though they are slower (Thomas et al., 2016). However, they do have difficulties with recognising double-digit numbers (Ansari et al., 2007). It is unclear whether difficulties to recognise double digits relate to their visuo-spatial abilities or whether with age and training digit recognition abilities can be improved. In sum, the limited number of studies that have examined arithmetic skills and that those that have included relatively easy problems, the true proficiency of people with WS in this area of mathematical development is currently unclear.

Predictors of mathematical abilities. Although a number of studies have compared performance in WS to mental aged matched groups, which allows insight into whether performance in WS is in line with their mental age abilities, few studies have directly examined what domain-general or other domain-specific abilities relate to performance on mathematical tasks in WS. In addition, reported information is sometimes insufficient to make judgements about what measures were used or specific findings.

Similar to typical development, mathematical abilities in WS have been argued to relate to estimation abilities, especially early in development (Rouselle et al., 2013). Rouselle and colleagues (2013) examined the acuity of large number, space, and time estimation in WS and found that number estimation related to acquisition of the first symbolic numerical competences. Other cross-sectional studies have shown that approximate non-verbal estimation abilities are in line with non-verbal abilities in WS (Rouselle, Dembour, & Noël, 2013; Van Herwegen, Ranzato, Karniloff-Smith & Simms, 2019). As it is possible that non-verbal abilities in WS do not exceed those of a 6-9-year-old child, estimation may remain relatively low in WS throughout development. Alternatively, it has been proposed that atypical attention patterns early on in life affect the development of non-verbal estimation abilities (Steele et al., 2012; Karmiloff-Smith et al., 2012).

Steele et al. (2012) established that early cardinality skills were a unique predictor of increased mathematical abilities in WS. Ansari and colleagues (2003) examined the contribution of verbal and visuo-spatial abilities in relation to cardinal knowledge. This study showed that whilst visuo-spatial abilities, and not verbal abilities, were the main predictor for mathematical abilities in the younger typically developing group, verbal abilities, but not visuo-spatial abilities, predicted performance in the WS group.

Together these studies suggest a developmental pathway with impaired attention and visuo-spatial abilities early on in life impacting the development of early domain-specific abilities, such as cardinality and estimation skills, which are important for the development of mathematical abilities later on in life (see Karmiloff-Smith et al., 2012; Van Herwegen, 2015). In addition, relative strengths in verbal skills, combined with general intellectual abilities, may provide (limited) compensation to overcome the impact of weak non-verbal skills. This interaction of skills may explain why studies have found that mathematical performance in WS is in line with verbal or overall intellectual abilities. For example, O'Hearn et al (2007) found that performance on TEMA in WS was higher than their overall visuo-spatial abilities and correlated to verbal and general reasoning abilities.

Methodological quality of the studies

Due to the rarity of the disorder, most of the studies included a relatively small number of participants. The average sample size across all of the different identified studies was 19.82 participants (SD = 13.19). However, the median was only 14 participants and just 5 studies included more than 30 participants. In addition, most of these studies included participants of a wide age range, with an average age range of 22 years and 6 months (SD= 16 years and 10 months) and median age range of 17 years.

There are many reporting issues within the identified papers. Five studies did not report the age range of participants, therefore it is difficult to compare data within these papers to others. Only one identified paper reported effect sizes (Steele et al., 2012). Many papers reported group means for outcomes without standard deviations (e.g., Patterson, 1999) so that it was not possible to calculate effect sizes.

Another methodological issue that was noted in the majority of studies relates to the control group(s) included in the studies. The use of either a chronological or mental age matched control group has both theoretical and practical implications (see Thomas et al., 2009 for a discussion). Regardless of the type of control group used, most studies argue that their group of individuals with WS and control groups are matched. Yet, the majority of these studies (with exception of Ansari et al., 2003) did not describe how the two groups were matched (i.e., were participants matched individually and if so what age or mental age score was considered a match?). In addition, these studies used frequentist statistics instead of Bayesian analyses to show that the groups did not differ in terms of the participant groups' chronological age or mental abilities. This is, of course, rather problematic.

A few studies have just included control groups that consist of younger typically developing children that are not matched in any way to the WS group (Ansari et al., 2007; Krajcsi et al., 2009; Libertus et al., 2014; Opfer & Martens, 2012; Steele et al., 2012) or have included no control group whatsoever (Van Herwegen et al., 2008). Although non-matched control groups can provide some insight into whether performance is in line with this control groups' age or ability range, the understanding that this methodological approach allows in relation to the cognitive mechanisms underpinning mathematical development in WS may be limited. This is due to development in this population often being delayed and also following atypical developmental pathways. Therefore, these studies are limited in explaining precisely why performance in WS differs from certain control groups as range of factors (e.g. age, overall ability, unmeasured skills etc) could account observed differences.

Discussion

The current review identified only 27 studies in 22 publications that have examined mathematical outcomes in individual with WS. This strongly contrasts with number of studies that have examined language development in this population (see review by Brock, 2007). Overall, the majority of studies have focused on whether mathematical abilities are spared (i.e., in line with chronologically aged matched controls or those of a similar overall cognitive abilities such as individuals with Down syndrome) or impaired (i.e., in line with younger typically developing children) in WS. About half of these studies used author-generated outcome measures. In addition, those studies that have used standardised measures have often included different standardised measures as well. The fact that very few studies have used the same measures means that synthesis of the mathematical abilities in WS is not possible from a mathematical point of view. However, most of the studies have shown that mathematical abilities in WS are impaired and that mathematical abilities do not exceed those of typically developing children aged 8 to 9 years old. Yet, very few studies have truly examined the development of mathematical abilities either using cross-sectional designs (e.g., Van Herwegen et al., in press) or longitudinally (Howlin et al., 2010; Steele et al., 2012; Udwin et al., 1996). In addition to the lack of studies that have examined what mathematical abilities relate to in WS (e.g., Ansari et al., 2007), there is very little known about the mechanisms that drive or hinder mathematical abilities in WS, **including the impact of domain-general difficulties**. This lack of research also explains why thus far, only one intervention study has been carried out to evaluate how mathematical abilities in WS can be improved.

Examination of the types of mathematical abilities that have been studied in WS shows that a number of studies have focused on previously identified “core mechanisms” of mathematical abilities, including small and large quantity symbolic and non-symbolic estimation abilities and counting skills. Although most of these studies have used different outcome measures and thus their results are not necessarily directly comparable, a number of conclusions can be drawn from the research.

It is very often reported that language is a strength in comparison to other abilities in WS (Jarrold, Baddeley, & Hewes, 1998). However, studies that have examined language abilities in more depth have shown that there is a gap between language production and comprehension and that individuals with WS are able to use words and language that they may not necessarily understand (Brock, 2007; Naylor & Van Herwegen, 2012). Similarly, studies examining mathematical abilities have shown that although people with WS often know the number names, cardinality or understanding the purpose of counting or knowing how many items are in a set when having counted the set of objects is problematic for people with WS. This finding has been replicated at least five times across studies using different ages and methodologies. This shows that counting skills and understanding of the counting principles are not really a strength for people with WS but counting abilities are delayed and may develop atypically (Ansari et al., 2003). Another conclusion that can be drawn from the research thus far is that, although people with WS are able to estimate small sets of objects, development of larger magnitude discrimination is impaired and remains low across development, probably below the age of a 9-year-old typically developing child (Rouselle et al., 2013, Van Herwegen et al., 2019). Although it is possible that magnitude discrimination difficulties link to a lack of understanding of how numbers relate to each other (Paterson, 2000) or impaired number line abilities (Opfer & Martens, 2012), it is unclear whether this difficulty links to more boarder domain general visuo-spatial or attention difficulties that may be present from infancy onwards (Karmiloff-Smith et al., 2012; Van Herwegen, 2015). A final conclusion that can be drawn from this review is that very few studies have focused on arithmetic development or how people with WS solve simple sums and that, although studies so far show arithmetic abilities are not in line with chronological age, the true arithmetical ability of people with WS is currently unknown.

Gaps in the literature

Examining the type of studies conducted thus far in this field also highlight a number of gaps within this area of research. Out of the 27 studies, 24 were conducted in either the UK or USA and only three were conducted in other countries (Belgium, Germany, and Hungary respectively). Seeing that the language of the number name system can impact on number processing in typically developing children (Göbel, Shaki & Fisher, 2011), the lack of cross-cultural research means that the global perspective of the development of mathematical skills in WS is relatively unknown and it is not clear whether the transparency of the number naming system impacts on the mathematical development of people with WS.

Despite the fact that the majority of studies have shown that mathematical abilities are delayed in individuals with WS, only one study (Opfer & Martens, 2012) thus far has examined whether mathematical abilities can be improved through specific mathematical interventions. Opfer & Martens (2012) provided feedback whilst participants completed a number line task. The study found that the participants with WS in the feedback group improved overall more than those in the non-feedback group (effect size of .37 pre-to-post in feedback group), even though there was no shift from logarithmic to linear function as a result of the training provided. This suggests that performance on a numerical task can be improved with a relatively simple intervention.

A number of studies mention that there is substantial individual variability (Krajcsi et al 2009; Thomas et al., 2006) with regards to the mathematical abilities in people with WS. However, very few of the studies have explored this individual variability in further detail (but see O'Hearn & Landau, 2007 who did item-by-item analysis and found that individuals with WS are not equally impaired on all types of TEMA items). As most studies have included participants of a wide age range, it is currently not clear whether this individual variability simply relates to development (either development in relation to chronological age or in terms of developmental age) or whether this variability relates to the disorder itself. Alternatively, it is not clear whether there might be different sub-groups based on the different mathematical difficulties that people with WS may experience just like there are sub-groups of children with mathematical difficulty or dyscalculia whose mathematical difficulties seem to relate to different underlying mechanisms (Bartelet, Ansari, Vaessen & Blomert, 2014; Costa, Nicholson, Donlan, & Van Herwegen, 2018).

Finally, studies so far have mainly examined particular domain specific mathematical abilities (i.e. cardinality, non-symbolic number abilities etc.). However, development is complex with small changes in development having cascading effects on other developmental pathways (Karmiloff-Smith, 1998). In addition, development is flexible in that due to plasticity alternative developmental pathways might be followed (Van Herwegen & Karmiloff-Smith, 2015). Yet, within the research in WS thus far, few studies have examined how these number difficulties relate to one another or how mathematical abilities relate to domain general abilities. For example, Steele and colleagues (2012) have shown that, although mathematical abilities in WS rely on verbal and non-verbal abilities, cardinality abilities contributed unique variance to the mathematical abilities of young children with WS. In addition, they showed that cardinality understanding related to sustained attention abilities. Ansari and colleagues (2003) showed that older children and adults with WS have issues with cardinality and that cardinality abilities related to verbal, rather than non-verbal, abilities. Therefore, these studies suggest that domain general abilities, such as impaired attention, might provide a causal explanation for relative mathematical strength and difficulties in WS (see Van Herwegen, 2015 for a discussion). Others have argued that people with WS might rely on their verbal abilities to overcome their mathematical difficulties but there might be limitations in how far verbal strategies can mediate mathematical difficulties (Steele et al., 2012).

Yet, most of these studies relied on cross-sectional data (with exception of Steele et al., 2012) and there is a lack of longitudinal studies that examine how domain general and domain specific abilities relate to mathematical development over development in WS. A recent study by Van Herwegen et al (2019) examined eye movement patterns during an non-symbolic estimation task and found that, although there were no differences in accuracy in the estimation task between the participants with WS, DS and the younger TD groups, those with WS and Down syndrome performed worse than the chronological age TD group. There were also no differences in looking patterns during the task as measured by the eye tracker. However, relationships between performance on the estimation task and looking behavior showed that older participants with WS had shorter looks, and switched more frequently, between the two sets of dots before making a decision. In addition, those with shorter looks performed better on the task. This suggests that looking behavior and task approach may change over development and this may impact on what abilities predict mathematical abilities in WS over development.

Future research on mathematical development in WS

Improving our understanding of mathematical abilities of people with WS does not only require additional research, a number of improvements can be made to research in this area in order to ensure that solid reliable conclusions can be drawn. First of all, a large number of studies have included a small number of participants with a wide age range. This is not surprising seeing the rarity of the disorder. However, future studies could look at sharing similar outcome measures or use a multiple lab approach (see for example, Van Herwegen, Purser, and Thomas, 2019) in order to create larger or even longitudinal data sets.

Secondly, in terms of reporting, none of the studies in the review reported any information about the socio-economic status (SES) background of the participants included in the study. This is problematic, especially for the control groups, as it has been shown that children from lower SES backgrounds are at a greater risk for mathematical difficulty (Morsanyi, van Bers, McCormack, & McGourty, 2018). It is therefore not always clear whether the control groups in the studies included typically developing children who perform within the typical range for mathematical abilities or whether these control children are likely to be at risk for mathematical difficulty as well. Therefore, future studies should include a number of details, including SES of the participants, details about the home language, the means, ranges, and standard deviations related to the ages of the participants, details of how participant groups have been matched and especially the standard deviations and effects sizes for the group means so that overall synthesise of the data will be possible in the future, In sum, although a number of studies have been published investigating the mathematical skills of people with WS, this area has been under-researched. A number of questions and research gaps in relation to the development of mathematical abilities in this population remain unanswered, especially **related to the cognitive mechanisms that may impact on mathematical development in WS, how these mechanisms may differ from typical development, as well as how mathematical abilities in WS can be improved.** The lack of research in this area also explains the lack of targeted interventions.

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Table 1: Summary of papers from systematic review

Author	N	Mean age (range or SD) *if included in paper	Mathem
Ansari, Donlan, & Karmiloff-Smith (2007)	WS= 31 • 18 children • 13 adults Control= 63 (4 groups) • 15 4-5 year-olds • 22 6-7 year-olds • 14 9-10 year olds • 12 adults	WS: • children: 9.7 yrs (SD= 1.9 yrs) • adults: 28.9 yrs (SD= 10.5 yrs) Control: • 4-5 year olds: 4.8 yrs (SD= .4 yrs) • 6-7 years olds: 6.9 yrs (SD= .6 yrs) • 9-10 year olds: 9.5 yrs (SD= .5 yrs) • 12 adults: 30.8 yrs (SD= 9.2 yrs)	Subitizi Digit rec Countin
Ansari, Donlan, Thomas, Ewing, Peen, & Karmiloff-Smith (2003)	WS= 14 NVMA= 14 NV & VMA= 14	WS= 7.2 yrs (6.0-11.5 yrs) NVMA= 3.5 yrs (3.0- 4.7 yrs) NV & VMA= 3.4 yrs (2.6 yrs- 5.3 yrs)	Countin Cardina
Dolscheid & Penke (2018)	WS= 10 DS= 15 NVMA= 10	WS= 12.6 yrs (6.16-16.8 yrs) DS= 10.9 yrs (6.6-16.7 yrs) NVMA= 5.0 yrs (4.1-6.1 yrs)	Cardina
Howlin, Davies, & Udwin (1998)	WS= 62 No control group	WS= 26.5 yrs (19.0-39.0 yrs)	Arithme

Howlin, Elison, Udwin & Stinton (2010)	WS= • 92 cross-sectional • 47 longitudinal No control group	Cross-sectional= 32.0 yrs (19.11- 55.4 yrs) Longitudinal= 36.10 yrs (25.7-49.7 yrs)	Arithme
Krajcsi, Lukács, Igács, Racsmány, & Pléh (2008)	WS=8 Controls= • Young=10 • Medium=10 • Older=10	WS= 17.8 yrs (12.0- 23.1 yrs) Controls= • Young= 8.5 yrs • Medium= 9.7 yrs • Older= 10.3 yrs	Additio Multipli Digit co
Lane, Van Herwegen, Freeth (2019)	WS= 24 Sotos syndrome= 20 TDs= 25	WS= 21.04 (8.52-52.25) Sotos syndrome= 18.43 (8.00- 37.42) TDs= 18.04 (7.92-42.08)	large no

Libertus, Feigenson, Halberda, Landau (2014)	EXPERIMENT 1: WS=18 Control=25 • 13 4-year-olds • 13 6-year-olds • 12 9-year-olds EXPERIMENT 2: WS=12 No control group	WS= 15.5 yrs (7.0-32 yrs) Control groups: • 4 year olds= 4.6 yrs • 6 year olds= 6.6 yrs • 9 year olds= 9.4 yrs WS= 19.9 yrs (12.0-29.0 yrs)	Large n Large n verbal e
O'Hearn & Landau (2007)	WS= 14 NVMA= 14	WS= 17.9 yrs (SD= 7.4 yrs) NVMA= 6.2 yrs (SD= 1.1 yrs)	TEMA-
O'Hearn, Hoffman, & Landau (2011)	WS= 15 Control = 42 • 14 4-year-olds • 14 5-year-olds • 14 6.5-years-olds	WS= 20.8 yrs (11.3-48.8 yrs) Control groups: • 4 year-olds: 4.6 yrs (4.0-4.11 yrs) • 5 year-olds: 5.7 yrs (5.0-6.0 yrs) • 6.5 year-olds: 7.4 yrs (6.6-7.8 yrs)	Subitizi Countin

Opfer & Martens (2012)	EXPERIMENT 1: WS= 30 • 15 children • 15 adults Controls= • 160 younger children, • 81 older children • 99 adults EXPERIMENT 2: WS= 30 • 15 children • 15 adults Controls= • 44 children in feedback group • 52 children in no-feedback group	WS= • children: 11.8 yrs (6-17 yrs) • adults: 33.7 yrs (18-56 yrs) Controls= • younger children: 7.9 yrs (6.4-8.9 yrs) • older children: 10.0 yrs (9.0-11.2 yrs) • adults: 22.3 yrs WS= • children: 11.8 yrs (6-17 yrs) • adults: 33.7 yrs (18-56 yrs) Controls= 7.9 yrs (6.4-8.9 yrs)	Number Number interven

Paterson, Girelli, Butterworth, & Karmiloff-Smith (2006)	<u>EXPERIMENT 1:</u> WS= 11 DS=18 MA=16 CA=14 <u>EXPERIMENT 2:</u> WS= 8 DS= 9 MA=8 CA=8 <u>EXPERIMENT 3:</u> WS= 8 DS= 7 MA=8 CA=8	WS= 2.5 yrs (2.0-3.0 yrs) CA= 2.5 yrs (2.0-3.0 yrs) MA= 1.3 yrs (1.0- 1.7 yrs) DS= 2.5 yrs (2.0-3.0 yrs) WS= 20.9 yrs (10.1-32.9 yrs) DS= 24.3 yrs (11.4-35.3 yrs) MA= 6.11 yrs (5.2-8.11 yrs) CA= 21.1 yrs (9.10-29.8 yrs) WS= 20.9 yrs (10.1-32.9 yrs) DS= 26.4 yrs (17.11-35.3 yrs) MA= 6.11 yrs (5.2-8.11 yrs) CA= 21.1 yrs (9.10-29.8 yrs)	Small n compari Large n Countin
Patterson (2000)	WS= 8 CA= 8 MA= 8 DS = 7	WS= 20.9 yrs (10.11-32.9 yrs)	Digit co
Robinson & Temple (2013)	WS= 1 DS= 1 TD= 21	WS= 15.9 yrs (N/A) TD= 9.5 yrs (SD= 4.03 yrs)	Definin Additio
Rousselle, Dembour, Noel (2013)	WS= 20 VMA= 20 NVMA= 20	WS= 22.1 yrs (5.6-52.1 yrs) VMA group: 7.6 yrs (4.6-11.8 yrs) NVMA group: 6.1 yrs (3.8-10.4 yrs)	Large n Cardina
Scerif, Ansari & Karmiloff-Smith (unpublished)	WS=14 CA= unclear VMA= unclear	WS= 25.9 yrs (SD= 3.11 yrs) CA= 25.6 yrs (SD= 4.1 yrs) VMA= 13.11 yrs (SD= 3.1 yrs)	Digit co

Steele A., Brown, & Scerif (2012)	<u>EXPERIMENT 1:</u> WS= 14 DS= 19 MA= 16 <u>EXPERIMENT 2:</u> N= 27 DS= 26 TD= 22	WS= 28.98 yrs DS= 29.11 yrs MA= 15.21 yrs WS= 6.7 yrs DS= 6.11 yrs TD= 3.5 yrs	Large n TEMA- Countin Cardina
Thomas et al. (2006)	WS= 16 CA=16 VMA=16	WS= 25.1 yrs (12- 53.0 yrs) CA= 30.4 yrs (12.2- 53.1 yrs) VMA=15.4 yrs (4.4-17.6 yrs)	Digit na
Udwin, Yule, & Martin (1987)	WS= 44 No control group	WS= 11.1 yrs (6.0-15.1 yrs)	Arithme R
Udwin, Davies, & Howlin (1996)	WS= 23 No control group	WS= 21.9 yrs (19.0-24.1 yrs) **Longitudinal cohort- first tested mean age = 12.1 yrs (10.1-15. 9 yrs) (Udwin et al., 1987)*	Arithme
Van Herwegen, Ansari, Xu, & Karmiloff-Smith (2008)	<u>EXPERIMENT 1 & 2:</u> WS= 9 No control group	WS= 2.11 yrs (1.1-4.5 yrs)	Small a discrimi
Van Herwegen, Ranzato, Karmiloff-Smith & Simms (2019)	WS= 24 CA= 24 MA= 24 DS= 23	WS= 19.4 yrs (8.0- 51.1 yrs) CA= 18.0 yrs (7.1- 42.0 yrs) MA= 6.1 yrs (4.1- 10.0 yrs) DS= 21.1 yrs (8.8- 49.2 yrs)	Large n

Note: WS= Williams Syndrome; DS= Down Syndrome; TD= typically developing; MA= mental age matched; CA= calendar age matched; VMA= verbal mental age matched; NVMA= non-verbal mental age matched

Figure 1.
PRISMA flow diagram illustrating study selection.