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The underreporting of vision problems in statutory documents of children with Williams syndrome and Down syndrome

Abstract

Vision problems can lead to negative developmental outcomes. Children with Williams syndrome and Down syndrome are at higher risk of vision problems, and these are less likely to be detected due to diagnostic overshadowing and difficulty accessing eye-care. Education, Health and Care (EHC) plans are statutory documents, introduced by the Children and Families Act 2014 in England, with the intention of integrating provision across these domains. Vision issues should be reported in these plans, and recommendations made about appropriate adjustments for them. We analysed the EHC plans from 53 children with Down or Williams syndrome. Our results showed significant underreporting, especially for children with Williams syndrome, and little explanation of what adjustments should be made. We also report pockets of good practice.

Keywords: Down syndrome; Williams syndrome; Special Educational Needs; vision; neurodevelopmental disorders

The underreporting of vision problems in statutory documents of children with neurodevelopmental disorders

Vision problems can have a negative impact on a range of life outcomes, including education, health, and social functioning (Davidson and Quinn 2011). The majority of learning in schools is based around visual tasks, meaning that children with visual difficulties are automatically at a disadvantage. This is especially the case for reading: text is typically printed in smaller font as the content becomes more difficult (Hughes and Wilkins 2000), so uncorrected vision problems can serve as a barrier to making progress. Other common vision problems may cause difficulty tracking moving objects, visual stress or fatigue making it uncomfortable to focus for long, and trouble with fine motor skills or eye-hand coordination; all of which may reduce classroom engagement (e.g. Schneck 1996). For many children, early detection and appropriate care can mitigate this disadvantage (Ma et al. 2014). However, this is less likely to happen for children with neurodevelopmental disorders, despite them being at higher risk of vision problems. This is because typical school-based vision screening is often not useful for this group; and they are less likely to access a sight test outside of school (Donaldson et al. 2019). There is also a risk of diagnostic overshadowing, with difficulties caused by vision loss assumed to be a result of their disorder (Carvill 2001).

Two neurodevelopmental disorders which are known to confer an increased risk for vision problems are Down syndrome (DS) and Williams syndrome (WS). DS affects around 1 in 1587 people in the UK (Alexander et al. 2016), and WS about 1 in 18000 (Williams Syndrome Foundation 2019). Almost 80% of people with DS are estimated to have vision issues (Krinsky-McHale et al. 2012), and at least 50% of children with WS are thought to have ocular problems (Atkinson et al. 2001). Common issues in both disorders are refractive error (requiring glasses) and strabismus (squint) (Woodhouse et al. 1997; Atkinson et al. 2001). People with WS are also at high risk of cortical, cerebral, and visual-motor visual impairments

(CVI), where abnormal brain development or brain damage results in visual processing problems. These range from difficulties perceiving motion or colour to effective blindness, despite typical eyes and optic tracts, but a relative lack of diagnostic specificity means that they are widely underdiagnosed (Philip and Dutton 2014). The total proportion of children with WS who have a vision problem is therefore likely substantially higher than 50%.

Despite similarly high rates of vision problems in both of these populations, information in the public domain about typical symptomatic profiles of each syndrome does not equally represent this risk. Whereas for DS the increased rate of vision problems is generally included in public information briefings (e.g. Down's Syndrome Association 2019), this information is missing from comparable profiles of WS (e.g. Williams Syndrome Foundation 2019). This suggests that awareness of vision problems in children with WS is likely to be low, even for groups who should be well-informed. Whilst teachers and other professionals, including teaching assistants and educational psychologists, report being relatively well-informed about DS (Lee et al. 2005), they are less likely to have detailed knowledge about the profile of less common conditions such as WS (Van Herwegen et al. 2019). This mirrors parents' concerns that professionals are poorly informed about WS and do not adequately tailor provision to their children's needs (Ashworth et al. 2019).

The importance of professionals being aware of the broader health and social needs of the children they work with was recognised in the introduction of Education, Health, and Care (EHC) plans in England 2014. The aim of the EHC plans was to achieve a more integrated approach to provision for children and young people with disabilities, ensuring that everyone involved in delivering services relating to a child's education, health, or care is aware of all other areas; facilitating a consistent approach. The EHC plan should cover personal information; the special educational, health, and social care needs of the child and provision required to address these; the outcomes sought, the type of placement required, and any

personal budget allocated. The required provision set out in the plan is legally required to be provided for the child or young person, up until the age of 25 years (Department for Education 2015).

However, recent analyses of the quality of EHC plans have suggested that they may not be delivering their intended aims (Castro et al. 2019). The systems for drawing up the EHC plans vary across Local Authorities, with no particular legislation mandating the consultation of particular professionals (Department for Education 2015). This risks some aspects of a child's needs being missed altogether.

In view of the high prevalence of vision issues in children with DS and WS, the current study reviewed the inclusion of information about children's vision status in their EHC plans. Our aim was to evidence whether they are currently being used effectively to provide information about appropriate adjustments and support needed e.g. in the classroom. Both conditions (DS and WS) confer a similarly increased risk of vision issues, but the literature suggests that this kind of syndrome-specific knowledge is less common both for parents and teachers of children with WS than those with DS. We therefore predicted that we would find poorer reporting of vision issues in the plans of children with WS. Our research questions were: 1) How many EHC plans make reference to vision issues?; 2) How does this relate to parental report of actual vision issues?; and 3) Are specific recommendations of adjustments for recorded vision issues made in EHC plans?

Methods

EHC plans were obtained for 53 children (see Table 1). These participants were recruited via social media and parental support groups. Contact details for a follow-up survey were available for 42 of the parents (23 of children with WS, 19 of children with DS). Responses were returned by 37 parents (response rate DS 89%, WS 87%). EHC plans were provided by parents and fully anonymised for analysis. A short follow-up survey was emailed

to parents asking whether their child had received a recent sight test, whether they were aware of any problems with their child's vision, and for details of any known problems. Face validity of this survey was confirmed by members of SeeAbility's Eye care and Vision team, who run a specialist eye examination service for children attending special schools. This study received ethical approval from the Research Ethics Committee at Kingston University.

Results

1) How many EHC plans made reference to vision issues? Overall, just under half (47%) of the EHC plans reported an issue with the child's vision. A Fisher's Exact test showed that this rate of reporting was significantly higher for the children with DS (70%) compared to those with WS (33.3%), $\chi^2 = 0.012$, $p < .05$. The rate observed for children with DS is therefore in line with previous reports based on actual optometric/ophthalmic assessments, whereas for children with WS it appears that vision issues are underreported (see Table 2).

2) How does this relate to parental report of actual vision issues? We received responses about 17 of the 20 children (85%) with DS, and 19 of the 33 children (58%) with WS. For the children with DS, all parents reported their child had a problem with their vision and 12 (71%) had a vision issue mentioned in their EHC plan. For the children with WS, 13 (68%) parents reported their child to have a problem with their vision of which just 4 (31%) had this vision issue mentioned in their EHC plan.

The most common issue for both groups was refractive error (12/13 of children with WS, and 14/17 of those with DS). Eight of the children with WS also had a squint, one had no binocular vision and colour vision deficiency, and one had a suspected CVI. Three children with DS had a nystagmus, one had a squint, three had a history of cataracts, and one used prisms to correct for an abnormal head position. This information is summarized in Table 3. Of those children with WS who had no mention of a vision problem in their EHC plan, one had suspected CVI, one had a squint, one had no binocular vision, one showed colour vision

deficiency, and one had been prescribed glasses for long-sightedness but was unwilling to wear them. This demonstrates that children with WS have a range of complex vision difficulties that are not reported in their EHC plans.

3) Are specific recommendations of adjustments for recorded vision issues made in EHC plans? Very few EHC plans made any specific recommendations for adjustments for recorded vision issues. For children with WS, none of the EHC plans made specific recommendations for any adaptations to be made for the child's vision problem. For children with DS, four plans (28%) noted only that the children needed to wear glasses, with no further information provided. However, there were some examples of good practice, with three plans (21%) making very specific recommendations for the classroom (see Supplementary Table 1). Only four plans included input from a vision professional.

Discussion

This study investigated how vision issues are reported in the statutory documents of children with WS and DS. There was clear evidence for underreporting of issues, and information about the functional impact of vision issues and practical adaptations that should be made for them were only included in half of those mentioning vision issues. Another important finding was that children with WS were less likely to have an existing vision issue noted in their plan than children with DS, despite these issues being known to parents.

Parental report of sight problems confirmed our hypothesis that there is true underreporting of vision issues in EHC plans, and that this is particularly the case for children with WS. For children whose parents responded to a question about actual visual status and reported a vision problem, less than a third had any mention of this problem in their plan. There are two reasons why children with WS in particular may be less likely to have their vision problems noted in an EHC plan: Firstly, WS is 12 times less common than DS, and correspondingly there is less research and public awareness. One recent survey of professionals

involved in the education of children with neurodevelopmental disorders (Van Herwegen et al. 2019) found that only 10% reported that the children with WS who they worked with had an issue with their vision; almost certainly a vast underestimation given what is known about the prevalence of vision problems in this population (Atkinson et al. 2001). The evidence provided by the current study showing worrying omissions of information around serious eye conditions for some children reinforces the need for better syndrome-specific training for professionals. Secondly, the range of vision problems associated with each syndrome is such that some of those associated with DS, especially refractive error, are also better understood and more easily detectable. In WS, a common visual comorbidity is CVI (Philip and Dutton 2014), which, despite being the foremost cause of childhood visual impairment, has no clear diagnostic and treatment pathways; resulting in a higher likelihood of diagnostic overshadowing for children with additional needs, such as WS (Van Den Broek et al. 2006).

One way in which the problems identified here could be addressed is by ensuring that vision specialists are always consulted when writing EHC plans. This could also help to improve detection rates of vision problems for children with neurodevelopmental disorders, by flagging children who had not received any eye-care. More comprehensive guidance over where and how issues such as sight problems should be reported in EHC plans would also help to improve reporting, ensuring that those preparing the reports were always prompted to enquire about a child's visual status, whether or not they were aware of the likelihood of raised prevalence in a particular group. Access to good examples of some adaptations to different vision conditions (such as those highlighted as good practice in Supplementary Table 1) may be useful to ensure that this advice is always provided.

The main limitation of this work is that we would ideally have been able to verify the actual visual status of all children reported on in this study via a full optometric assessment; rather than relying on parental report. Future work could therefore aim to carry out a more

comprehensive study to include such an assessment. It may also be of interest to compare these groups to typically developing children with visual problems only (i.e. no underlying developmental disorder) and EHC plans. Finally, given the wide variation in how vision problems are reported and not reported in EHC plans in this work, it may be interesting to look in more detail at facilitating and limiting factors of local resource provision underlying this variation.

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