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Review of Hypermobility and Ehlers-Danlos Syndrome type 3 from the safeguarding prospective

Case Study

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A 6-year-old boy is referred for community paediatric assessment with frequent leg pains which are reported as being worse following strenuous activity. The child was also then complaining of extreme lethargy and is missing a substantial amount of schooling due to this. The child is also reported to have frequent abdominal pains with some associated symptoms suggestive of an element of constipation but not significant to enough to explain the frequency of the pain. He is overweight but reported by the family to be picky with his diet.

Examining the child no abnormal signs are elicited. He is of normal appearance with no abnormal stigmata and normal skin elasticity. General examination is normal. The abdomen is soft and non-tender with no palpable faecal masses. Neurological examination is also normal with a normal gait, normal muscle bulk and normal reflexes. The muscle tone is possible a little low with relatively flexible joints but not significantly outside what you would expect to find in a child of this age.

Mum then reports that she has a diagnosis of Ehlers-Danlos (EDS) given following a private rheumatology appointment as does her older daughter who is now home educated as she was unable to cope with the physical demands made from her school, was unable to get out of bed in time for school on regular occasions due to her significant lethargy and also now has access to a wheelchair to allow her to be transported across longer distances.

Mum also then informs you that she is certain that her son has also acquired Ehlers-Danlos. She asks that you write to school confirming the diagnosis and explaining that he needs to be excluded from all physical activity as this causes too much tiredness and leg pains and often means that he cannot make it into school the following day. She also asks for a referral to the wheelchair service to help the family when walking around town or other longer distances. She also explains that she is now administering Calpol regularly four times a day for both leg and abdominal pains and asks you to request that school administer the lunchtime dose and for the GP to place this on his repeat prescriptions for medical reasons. She also volunteers that she is highly likely to be considering also removing the child from school at some stage in the future as she has no confidence that the school will be able to cope with his complex needs, as they were unable to cope with his older sister.

You have some limited knowledge of Ehlers-Danlos from medical school days and venture the opinion that you are not convinced that this is the case due to the absence of skin laxity. Mum looks at you witheringly that she is referring to Ehlers-Danlos type 3 which does not have the associated skin presentation.

You then go on to explain that you feel it is important to rule out other potential causes of the symptoms described and, at the same time you would look into checking a genetic profile looking for the abnormalities associated with Ehlers-Danlos. Once again mum responds with a level of contempt informing you that they have not yet identified the genetic abnormality associated with EDS type 3. She expresses exacerbation as to how little doctors know and understand about Ehlers-Danlos Type 3 and directs you towards the Ehlers-Danlos support group website telling you that it holds all the information you will need. You suggest a referral to your local paediatric rheumatologist but she declines saying that the diagnosis is already clear and all she needs is the support in putting into place the steps to prevent deterioration in his symptoms as well as referral to physiotherapy requesting an appropriate physiotherapy programme is put in place a well as referral to OT for support with his hand strength and to provide the equipment he will require.

All subsequent blood tests are normal including renal, liver and thyroid function, bone profile, inflammatory markers and creatinine kinase. Where next?

Communication with school complicates things still further. They report that his attendance is already poor and they have requested the input of the educational welfare officer. When the child does get into school they report that he can appear a little reluctant to separate from mum still but, once he is in the classroom they report him to enjoy his time at school and belts around the playground with all of his friends with no signs of pain or distress. He lacks a little confidence and will sometimes appear a little clumsy when running but this they feel that if they can get him into school more regular they could work with him around this. They feel that his issues are a result of his lack of normal childhood experiences.

When they have broached this with mum she has responded saying that she was not at surprised to hear that he was running around the playground as she experienced the consequences of this the same night when his leg pains were worse, causing him great difficulty in getting to sleep and then he was so much more lethargic the following morning that she could not then get him into school. She asked school to curtail such activities in future to prevent the deterioration she subsequently sees at home. She has told them that her child has this syndrome and she was about to get confirmation following the referral to the paediatrician who would then be able to support her with these requests.

School then raised the potential for fabricated and induced illness (FII) and have asked whether you would consider a referral to the social care team to ask them to assess further.

Discussion

Many of us within paediatrics will have encountered a potentially similar scenario as above. It may involve the raising of a potential diagnosis of Ehlers-Danlos type 3 or the wider concept of general hypermobility and its effects on day to day functioning. I suspect it will provoke mixed emotions in all of us. Being the empathetic paediatricians that I know we all are you will want to support and advocate for this child and their family but, equally you will have concerns as to what this child is missing out on within school, whether they are being unnecessarily restricted in what they can do and the potential for the move to home schooling will concern us, removing the one other regular set off professionals involved who can support the child and alert the local authority of any further worrying features that they observe.

So, what is the evidence base behind both hypermobility and Ehlers-Danlos, and specifically type 3?

Continues overleaf

Hypermobility

There is a general consensus of opinion certainly amongst paediatric rheumatologists that hypermobility is over diagnosed. The first point to stress is that there is no clinical definition of hypermobility. You may here some quote the use of the **Beighton score.** This is an assessment scale used in adults to assess range of joint movement. It is important to stress that it has never been validated in children and, even with adults it was validated in a very different South African population. Studies have been attempted to research use of the score in children using what could only be described as significantly floored observational research.
Joint mobility changes substantially both with age and between sexes. Normal variation is huge. Accepted research has noted an association with 'growing pains' or recurrent nocturnal limb pain in children and relative pes planus (flat feet). But the evidence around wider hypermobility and limb pain is not demonstrated. An association between hypermobility and learning disability also does appear to have level of acceptance but the communication difficulties present within this group make further studies significantly more difficult.

Interventions:

The critical factor of managing this group of children is preventing the child from becoming disabled by the condition. Sometimes pure reassurance around the range of normal mobility and the fact that increased actively will not lead to deterioration for the family to allow appropriate participation in childhood activities once more. In fact, providing the information that improving muscle strength by encouraging activity will in the long run, lead to less pain can be helpful.

The British Society for Paediatric and Adolescent Rheumatology has produced some excellence guidance around this issue obtainable through the following link: ***http://www.mcns.scot.nhs.uk/sparn/wp-content/uploads/sites/8/2017/01/Guidelines-for-Management-of-Joint-Hypermobility-Syndrome-v1.1-June-2013.pdf***

One problem arises, as is described in the above scenario, where the family have already learned themselves to be disabled by the condition. A parent has allowed the condition to limit what they themselves can do and see this as the only way forward with their child. This then leads to the question being raised around by those working with the family around the potential for FII a matter we will return to later.

Ehlers-Danlos Syndrome

I aim to focus on Type 3 within this article but, for completeness will comment on the other forms.
There have now been 13 types of EDS described. Type 1 is classical and Type 2 a variation of classical. They both present with very pronounced hypermobility and clear laxity of skin. The skin also has a very evident velvety texture. They will often present at around 9 months of age with abnormal amounts of bruising that can initially lead to understandable safeguarding concerns. The features described above though should fairly quickly lead to the correct diagnosis. The underlying genetic abnormally can also be identified through genetic analysis.

Type 4 (vascular type) is a tragic condition with extremely fragile arteries the invariable rupture leading to early death in childhood. It is an autosomal dominant condition so acquired through new mutation and there are no useful interventions presently available. Most of the other recognised EDS types are associated with differing forms of skeletal dysplasia resulting in severe levels of mechanical pain needing proactive intervention. They are also often associated with skin laxity and abnormal bruising. They all have well defined genetic markers.

Name of EDS Subtype	IP*	Genetic Basis	Protein Involved
Classical EDS (cEDS)	AD	Major: *COL5A1, COL5A2*	Type V collagen
		Rare: *COL1A1* c.934C>T, p.(Arg312Cys)	Type I collagen
Classical-like EDS (clEDS)	AR	*TNXB*	Tenascin XB
Cardiac-valvular EDS (cvEDS)	AR	*COL1A2* Biallelic mutations that lead to *COL1A2* NMD and absence of pro α2(I) collagen chains)	Type I collagen
Vascular EDS (vEDS)	AD	Major: *COL3A1*	Type III collagen
		Rare: *COL1A1* c.934C>T, p.(Arg312Cys) c.1720C>T, p.(Arg574Cys) c.3227C>T, p.(Arg1076Cys)	Type I collagen
Hypermobile EDS (hEDS)	AD	Unknown	Unknown
Arthrochalasia EDS (aEDS)	AD	*COL1A1, COL1A2*	Type I collagen
Dermatosparaxis EDS (dEDS)	AR	*ADAMTS2*	ADAMTS-2
Kyphoscoliotic EDS (kEDS)	AR	*PLOD1*	LH1
		FKBP14	FKBP22
Brittle cornea syndrome (BCS)	AR	*ZNF469*	ZNF469
		PRDM5	PRDM5
Spondylodysplastic EDS (spEDS)	AR	*B4GALT7*	B4GalT7
		B3GALT6	B3GalT6
		SLC39A13	ZIP13
Musculocontractural EDS (mcEDS)	AR	*CHST14*	D4ST1
		DSE	DSE
Myopathic EDS (mEDS)	AD or AR	*COL12A1*	Type XII collagen
Periodontal EDS (pEDS)	AD	*C1R*	C1r

It must be stressed at this point that having a diagnosis of EDS quite clearly does not exclude you from the potential for child abuse. A child with EDS will potentially present with an abnormal amount and frequency of bruising but the bruising should still have an acceptable causation provided and, if not in an area where normal bruises would occur (shins, elbows and other bony prominences for example) then further investigation should take place to rule out the potential for the bruises being of non-accidental causation.

Ehlers-Danlos type 3 – Hypermobility variation

As suggested above, this is the one type where significant controversy remains. It presents with a subjective description of hypermobility (the objectivity of such a diagnosis does not exist as described above), limb pains, often abdominal pain and often with a family history.

Children present in 3 ways:

1. Symptomless but with the reported hypermobility and the family history of EDS type 3
2. Musculoskeletal pain with hypermobility
3. Already severely disabled with their condition with crutches/ wheel chair and often with associated idiopathic abdominal pain with the features that would be described in irritable bowel syndrome.

Unlike all of the other defined EDS types there is no genetic abnormality identified in this form. When providing the label, the clinician will be giving the diagnosis based on the subjective symptoms and signs listed. It is fair to say that the vast majority of paediatric rheumatologists would like to see type 3 removed from the EDS subtypes.

The Ehlers-Danlos support group website has taken the opposite position and is very pro-actively supportive of the diagnosis, advocates from the families around the diagnosis and promotes its on-going use despite the absence of medical evidence.
This leaves us as paediatricians in a dilemma when considering our approach when supporting a family where this diagnosis exists. At

present time, and with the diagnosis still very much in existence, it would not feel appropriate to be taking an extreme approach in not accepting that diagnosis from the family.

My suggestion around further management would therefore be the following.

A child presenting with a diagnosis of asymptomatic EDS type 3 is usually the easiest to manage. Reassurance to both the child and parents of the benign nature of observed hypermobility and describing how continuing normal daily activities will not lead to development of unpleasant symptoms is often enough.

For those who are already being described as being in pain by their family members the same approach of advising against any restriction in activity and encouraging regular school attendance should still be followed.

If this does not succeed or for those with already significant disability demonstrated a more proactive and multidisciplinary approach should be taken. Group sessions for children with hypermobility have been shown to be effective in enabling individuals to develop appropriate coping mechanisms. Specific orthotics have also been shown to demonstrate some positive effects specifically with those with pes planus though whether this is part placebo effect has been suggested. For those who develop hand pain with writing the use of a fatter pen of specific pen grips can be useful.

Safeguarding concerns

As hopefully demonstrated through the initial case study, concerns around safeguarding issues and specifically FII but also general emotional abuse can be raised within this area frequently. In the majority of cases, if a supportive and sympathetic approach is taken, observed hypermobility will not lead to the need for safeguarding interventions and the child can be rehabilitated into full participation. If this approach is unsuccessful though and a full multidisciplinary team approach does not produce a positive outcome and, after thoughtful discussion, there is felt to be the potential for a child being prevented by the action of their parents to participate appropriately in life activities, then referral for consideration of a section 47 assessment should be made. This quite evidently will not be an easy path to take though. The present strong advocacy around the existence and reported consequences of hypermobility and EDS type 3 will continue to make the process of proving that FII is occurring difficult. A strongly word statement from the British Society for Rheumatology around the existence of EDS type 3 as being a true subtype of the syndrome or not would clearly help.

The case study also brings up the issue around home schooling at the potential impact of this practice on identifying when child protection issues are occurring, but that is clearly a complex issue and one for a future debate.

Written with the kind support of Dr Clive Ryder – Consultant Paediatric Rheumatologist, Birmingham Children's Hospital

Children in care service improvement: action

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A review of the health pathway and provision of statutory health services for children in care in Nottinghamshire took place in 2016, which resulted in a multi-agency improvement programme. We hope to share the learning from this process, which is ongoing, and enable you to transcribe it to any change or service improvement project. The first article looked at general principles and planning, the second article reviewed setting expectations and concentrated on some of the specific needs of children in care.

This article will review a specific project and process; to review how health assessments for children in care are managed when those children and young people are placed in another area. The first acronym which may be unfamiliar to some is OOA, which stands for 'out of area' and relates to the health assessment but also the child or young person who is placed outside of our local Nottinghamshire area. We will also describe OLAC, which stands for 'other local authority children', so those placed in our area from anyway in the UK.

Context

There are many reasons a looked after child may live away from their home authority. Some need to be out of area to help keep them safe from dangerous influences closer to home. Others need the kind of specialist support that is not available in all local authority areas. Many children require long-term foster placements that are not available close to home. Some looked after children move out of area so that they can live with brothers and sisters, or to be cared for by relatives who are approved as foster carers (Ofsted 2014).

In comparison with the wider looked after children population, those placed out of area are likely to have experienced more placement moves and they are more likely to have mental health difficulties. They are more likely to go missing, and to be subject to the serious risks associated with going missing. Those with the most complex needs and challenging behaviour are more likely to be living in children's homes and young people living in children's homes are nearly three times more likely to be living outside of their local authority boundary and more than 20 miles from home than children living with foster carers (Ofsted 2014).

Statutory guidance (DH, 2015), makes it clear that when accommodation is arranged in the area of another CCG, the 'originating' CCG remains the responsible CCG for the services that CCGs have responsibility for commissioning, including the child's statutory health assessments. NHSE guidance (NHSE, 2013), also states that arrangements should be put in place to track looked after children that are placed out of area to ensure their needs are being met.

Background

A review of the health pathway provision of statutory health services for CIC in Nottinghamshire took place in 2016 (Hamilton, G). This piece of work identified three groups within our looked after children cohort who were at risk of poorer outcomes because of inequity within the commissioned service provision and these included

- Nottinghamshire children who are placed out of the county
- Other local authorities' children who are placed into the county
- Young people leaving care

At this time all out of area health assessments were organised and overseen by the health provider organisations internally. There was no formal involvement by the CCGs.

At that time the Designated Nurse for looked after children was based in a provider organisation; the post moved to the CCGs shortly after the publication of the Pathway review (Hamilton,