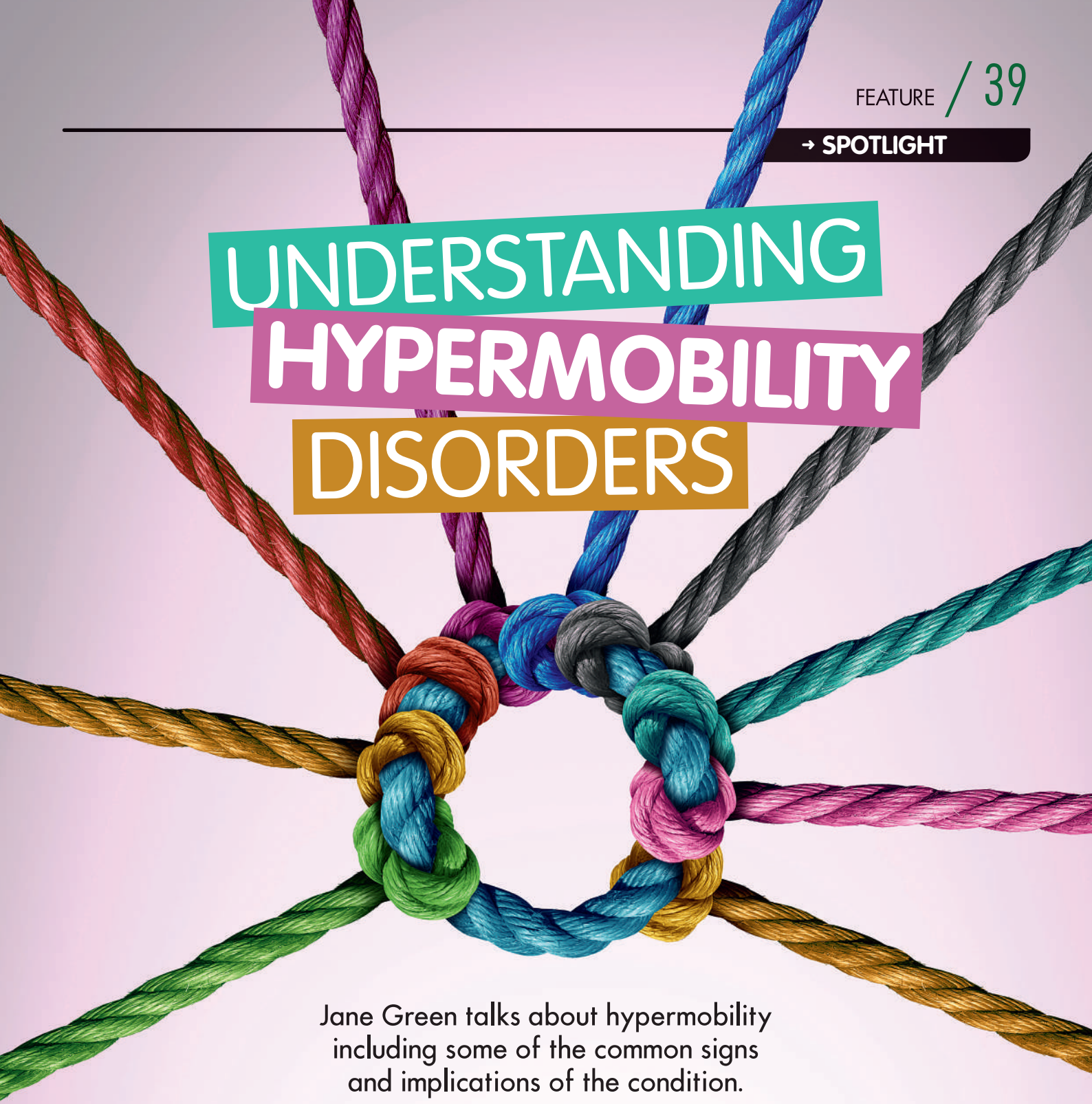
UNDERSTANDING HYPERMOBILITY DISORDERS



Jane Green talks about hypermobility including some of the common signs and implications of the condition.

Although diagnoses of Ehlers-Danlos syndromes (EDS) and the closely related Hypermobility Spectrum Disorders (HSD) are increasing amongst children and young people, little is known about the impact this has on their attainment and attendance in school. Emotional Based School Avoidance (EBSA) can occur as a result of EDS and HSD, but often emotional anxiety and/or developmental co-ordination disorders (DCD) or neurodevelopmental conditions, such as autism, are identified as the cause.

WHAT ARE THE HYPERMOBILITY SYNDROMES?

EDS (previously joint hypermobility syndrome) and HSD are heritable disorders that affect the connective tissues found throughout the body. People with EDS/HSD have defective connective tissues, but everyone presents differently, from being asymptomatic to severely disabled. Connective tissues can be thought of as cement between bricks holding up a house; some of the cement is in the right proportions, some not, and can be too weak or crumbly to support the building. If the cement is weak that is likely to affect other parts of the building like the electrics and waterworks.

There are around 14 types of EDS, the most common being hypermobility EDS. There are genetic tests for 13 but not for the most common termed hypermobility EDS which is diagnosed by clinical examination and gathering of the individual's history.

→ SPOTLIGHT

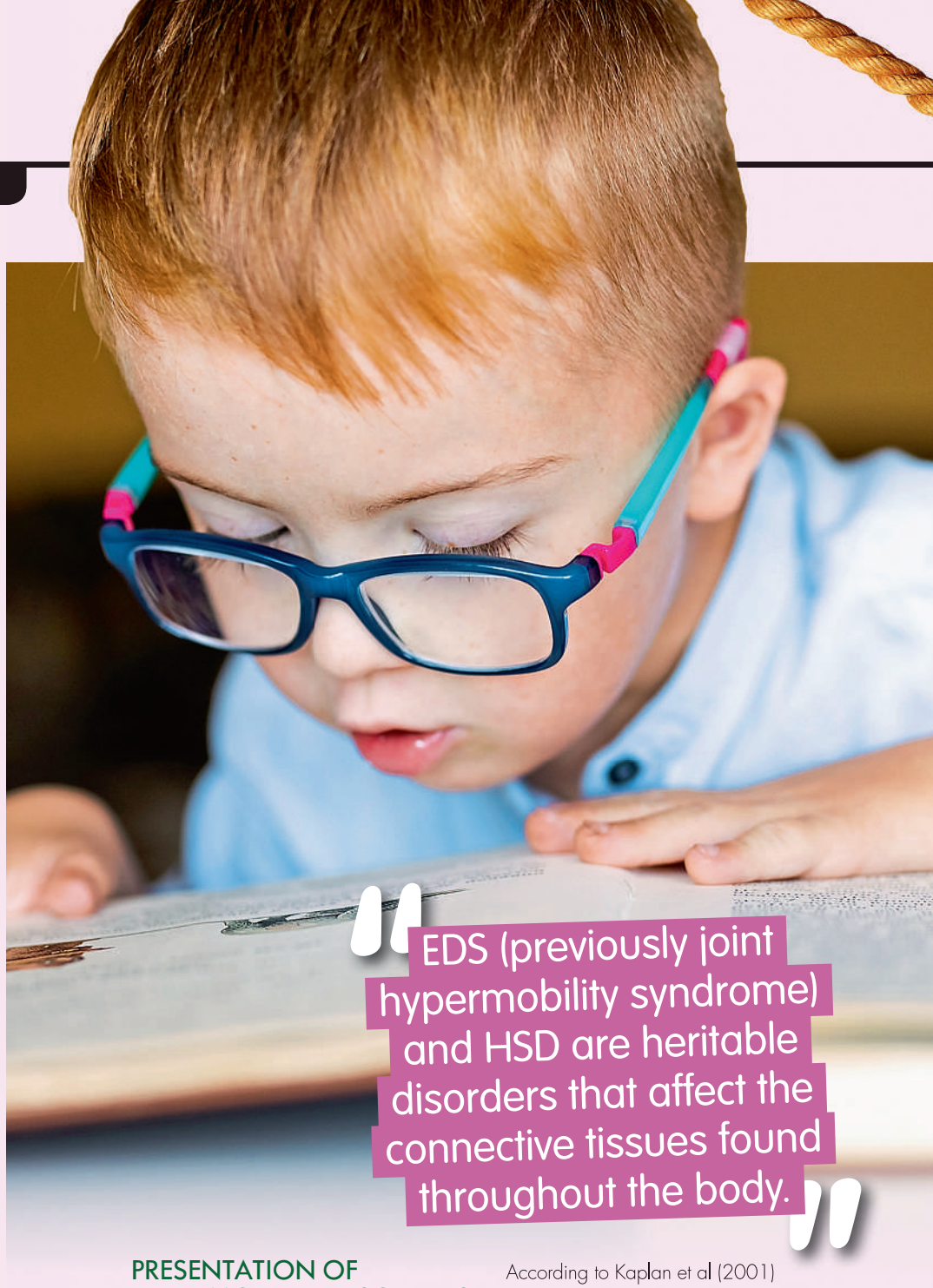
EDS was first described by Hippocrates in 400 BC as a condition with joint laxity and multiple scars (Parapiam, L. A. 2008). The syndrome is named after Edvard Ehlers (1863-1937) and Henri Alexandre Danlos (1844-1912). Subtypes of EDS were redefined three times in 1988, 1997 and 2017 (Jovin, 2020) from 13 to 14 subtypes. Progress in the management of EDS/HSD has been slow as there is no diagnostic care pathway. It is multi-systemic and can be complex.

It is considered rare as rates are 1/5,000 although some believe it is more likely to be 1/500 but undiagnosed (Demmler J et al, 2019 and Eccles J, 2020) possibly due to the unique complexity of the disorders, uncommon representation of symptoms, leading to psychosomatic assumptions and the very 'long' history. To this effect, the 'godfather' of EDS, Prof. R Grahame, has stated that 'no other disease in the history of modern medicine has been neglected in such a way as Ehlers-Danlos syndrome'.

PRESENTATION OF HYPERMOBILITY DISORDERS

Representations of being 'double jointed' are called to mind, however the effects of EDS/HSD are much broader and can impact on CYP's physical, emotional and psychological development. They might be able to extend their limbs much further than the average range, often dislocating, subluxing (half dislocating but 'going back in') or spraining without previous trauma and sometimes do not present as acutely in pain. Other symptoms are:

- extreme tiredness
- reflux
- heartburn
- stomach pain and spasms not linked to anxiety
- gastrointestinal issues
- vascular issues
- muscle tightness
- dizziness/clumsiness
- temperature issues
- neurological issues
- acute, persistent pain
- autonomic dysfunction of the autonomic nervous system (ANS) seen as palpitations/anxiety.



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PRESENTATION OF HYPERMOBILITY DISORDERS

Drops in blood pressure can cause 'brain fog'. Pupils can get surges of adrenaline that raise cortisol levels particularly in the co-occurring condition Postural Tachycardia Syndrome where an abnormal increase in heart rate after standing up can trigger experiences of anxiety; sensitivity to the chemical norepinephrine causes further anxiety. A study by Eccles (2020) suggests an increased rate of joint hypermobility in related neurodevelopmental conditions is linked to ANS dysautonomia and pain. Other difficulties are:

- Finger grip issues
- Poor swallow reflex
- Gastrointestinal reflex disease
- Fainting
- Headaches/migraine.

According to Kaplan et al (2001) 'overlapping difficulties in two or more developmental and educational domains implies that discrete diagnosis of a single disorder is often not appropriate'. A study by Clinch et al (2011), found the positive association of generalised joint hypermobility in adolescent females is at 45%. EDS/HSD injuries can occur such as wounds, bruising, dislocations, fractures and sprains without any preceding trauma.

INTEROCEPTION

Interoception, one of the lesser known senses, provides internal information about physical health (Quadt et al, 2018), mental health (Craig, 2015) and emotional functioning (Critchley and Garfinkel, 2017). It also relates to hunger, pain, thirst, digestion, heart rate, temperature,

itching, pain or pleasure. Often individuals with EDS/HSD are unaware they are hungry until ravenous. With co-occurring diagnoses, more confusing emotions may not be 'felt' and may be harder to identify. Individuals may not recognise fear because they do not recognise that tense muscles, shallow breathing, and a racing heart signal fear (Quadt, 2018).

ANXIETY AND ALLERGIES – INTOLERANCE OR ILLNESS?

Individuals with EDS/HSD often have food related allergies which cause anaphylactic reactions. Some are not diagnosed with, but some are, Mast Cell Activation syndrome (MCAS) which includes breathing issues, swelling, low blood pressure and diarrhoea. As a new disease there are no current NICE or NHS guidelines.

ATTENDANCE AND ATTAINMENT AND EMOTIONAL BASED SCHOOL AVOIDANCE

According to Pellegrini (2007), EBSA stands for Emotional Based School Avoidance (EBSA). The Government measures all non-attendance in one statistic called 'Persistent absence'. EBSA is a heterogeneous concept (Maynard et al, 2015) and cannot be treated as a single condition.

According to Harrowell et al (2017), 'Social communication differences and hyperactivity can have an effect on psychological outcomes'. If diagnosed with DCD, 'the evaluation and management of a child with suspected DCD, consideration of other possible co-occurring difficulties is essential.'

Not all Children and Young People (CYP) on the SEND register are attending or attaining in school due to what is first perceived as emotional anxiety. It might be considered to be this, but when physical, mental and emotional or social wellbeing needs are present, then EDS/HSD and other neurodevelopmental areas should be investigated further. Previous research on EBSA mentions somatic pain, headaches, stomach aches, poor digestion, bowel issues, tiredness, muscle rigidity and a racing heart as symptoms of anxiety due to emotional anxiety (Oxford, Coventry, West Sussex CC). Although this might be correct for some, the physical causes of this are not considered.

Outpatients can be a struggle for parents of EDS/HSD CYP outside school hours. Some children and young people manage most lessons but have no more energy for any extracurricular activity. Reasonable adjustments to timetables are not made, leading to injuries, extreme tiredness or anxiety, plus low self-confidence. Without multidisciplinary meetings, the dots are sometimes not joined up. Occasionally, parents/carers are positioned as 'the problem' due to a lack of awareness and knowledge that EDS/HSD are heritable multi-systemic disorders.

> "Until these disorders are widely recognised, children and young people will continue to be misunderstood and not attain or achieve in education."

BIO

JANE GREEN

Jane was the lead in autism education for leading autism charities leading the social model of autism education, an advisory consultant as well as Assistant Headteacher. She worked in most phases of education from EYS, primary, secondary, FE and special. She was however labelled 'dim' and at age 16 only gained two qualifications. She has two adult children and is late diagnosed autistic and has hypermobility EDS.