The interrelationship between Down syndrome, Autism and Joint Hypermobility within the dance teaching and learning environment

What is Joint Hypermobility?

Joint hypermobility (JH) is a heritable hypermobility spectrum of disorder (HSD) that includes generalised joint hypermobility (GJH), localised joint hypermobility (LJH), peripheral joint hypermobility and historical joint hypermobility. It is commonly associated with Marfan syndrome (MFS), Osteogenesis Imperfecta (OI) and Ehlers-Danlos syndromes (Castori et al., 2017).

Is Joint hypermobility associated with Down Syndrome and Austistic Spectrum Disorder?

At present there is increasing evidence and debate to suggest that there are associations between JH and both Down syndrome (DS)and Autism spectrum disorders (ASD) (Foley & Killeen, 2019; Talbot & Alshryda, 2017). There are a range of musculoskeletal complications for example, hip instability, patellofemoral instability (Talbot & Alshryda, 2017) and a combination of ligamentous laxity and low muscle tone (Foley & Killeen, 2019), that associate DS with JH. These complications, however, are secondary in DS with the result they are infrequently reported (Foley & Killeen, 2019). Such complications can, however, contribute to increased health risk of injury and a number of musculoskeletal disorders that potentially lead to a delay in acquisition of motor milestones (Przymuszała et al., 2018). Similarly, and whilst commonly only associated with stereotyped behaviours (such as social and communication problems along with other behavioural issues), ASD also presents with physical abnormalities that are also common in JH. These include the delay in gross motor and fine motor milestones, hypotonia and increased joint laxity (Anujot, Devi, & Swati, 2019). Autism spectrum disorder (ASD) and Down syndrome (DS) appear then to be interconnected with JH. This is not, however, always flagged up in the literature because each (DS, ADS & JH) has a very separate blanket term that describes the aetiologically and seemingly clinically unrelated pathologies that usually appear with these disorders in childhood. Moreover, DS, ADS & JH are typically seen by different medical fields, such as psychiatry in the case of ASD, specialist doctors for genetics and delayed intellectual development for DS and musculoskeletal disciplines and rheumatology in the case of JH related disorders. Thus, links between these pathologies are rarely established in clinical settings.

How does JH present in , DS and ASD?

Evidence of links between JH and altered brain development and activity in children, such as attention deficit disorders ADD, attention deficit hyperactivity disorders ADHD, Autism spectrum disorders ASD, clumsiness, poor coordination, proprioception and visio-spatial skills are increasingly presented as potential mechanisms through which JH and psychopathological symptoms interrelate (Sinibaldi, Ursini, & Castori, 2015). The need for research in this emerging area was recently highlighted in order to combine and better understand the somatic and developmental/psychiatric aspects of JH (Baeza-Velasco, Grahame, & Bravo, 2017). Associations between JH and ASD are indeed beginning to manifest in the literature and there is currently a growing body of research that suggests that ASD and JH can co-occur and the likelihood of this happening is more often than expected by chance (Baeza Velasco, Hamonet, Baghdadli, & Brissot, 2016; Eccles, Owens, Mathias, Umeda, & Critchley, 2015). Similarly JH is now becoming more frequently discussed with regards to

DS, however the current scoring systems for JH (e.g. Beighton scoring) fail to identify the hypermobile joints and the severity of hypermobility characteristics that are specific within DS (Foley & Killeen, 2019).

For the purpose of teaching dance in inclusive situations, it is clearly important, then, for teachers to understand the implications of JH in addition to understanding DS and ADS.

Joint Hypermobility within the environment for dance.

The implications of Joint Hypermobility are better understood from a bio-psycho-social perspective which is further explained here

Bio-markers of JH that are related to dance

At a biological level JH enables an extreme range of motion in the joints and manifests, for example, as hyperextension of the limbs. It is often characterised in lay terms as 'double-jointedness' and those who present with JH are also often described as having 'bendy bodies'. These characteristics are also very conducive to the physical demands and aesthetic that is coveted within dance and some aesthetic sports. Not surprisingly, then, JH is reported to be prevalent within dancers and commonly signposted as a 'potential asset' in vocational dance settings (McCormack, Briggs, Hakim, Grahame & Grahame, 2004; Scheper et al., 2013).

In addition to the excessive flexibility that is available in JH, teachers of dance need to also be aware of the less apparent signs and symptoms of JH. Joint hypermobility may indeed present as a symptomless trait (Castori et al., 2017) that is an asset to any dancer. However, the unwarranted use of the excessive range of motion within an inherently fragile hypermobile joint can lead to micro and macro trauma. Micro trauma is, for example, a silent recurring injury that is often not detected by the clinician or individual at the time it occurs (Castori et al., 2017). With overuse and repetitive movement, which is often inevitable in dance, this may lead eventually to pain, chronic pain, and bone degeneration. When specific, for example to the hip joint, a labral tear (this is a tear to the fibrocartilage, attached to the rim of the acetabulum, that helps keep the head of the femur in place) can occur (Devitt, Smith, Stapf, Tacey, & O'Donnell, 2017). Similarly, macro traumas, for example, dislocations, tendon ruptures and soft tissue injury, that occur due to instability and/or weakness of the joint, fragility of the connective tissue and also as a result of the overuse of an excessive range of motion that is often seen in dance (Castori et al., 2017).

Poor proprioception (sense of the body in space/body awareness) is also commonly associated with JH (Chopra et al., 2017; Fatoye, Palmer, Macmillan, Rowe, & van der Linden, 2009; Ghibellini, Brancati, & Castori, 2015; Nagai, Schilaty, Strauss, Crowley, & Hewett, 2018) and proprioception may also be further diminished as a result of the type of micro trauma discussed above. Reduced proprioception and muscle strength are of course interrelated; the one affecting the other in a vicious circle. Unfortunately this in turn limits the use of the very activities that are recommended for rehabilitation; to strengthen and return to healthy functioning in joints (Scheper et al., 2013, 2016). Diminished proprioception may also be the cause of the apparent clumsiness that has, in the past, been anecdotally associated with children with JH and more so in the literature over the past decade (Ghibellini et al., 2015). Indeed, clumsiness or delay in walking is often a silent precursory sign to the existence of JH (Adib, Davies, Grahame, Woo, & Murray, 2005; Ghibellini et al., 2015) and may then potentially also be a precursor to ASD, however this has yet to be substantiated in the literature. Clumsiness is often a warning sign to parents and paediatric practitioners that something may not be quite right and it is also often the motive for them to prescribe cautious physical activities (such as dance) as a means to improve the lack of coordination and/or delayed mature gait pattern. As dance is often prescribed in these situations, it is, therefore,

important that dance teachers are aware, not only of the clumsiness, but also the potential underlying condition of JH.

Another symptom that regularly features with JH is the apparent rapid fatiguing and deconditioning of the body (Keer & Grahame, 2003). It is important for dance teachers to understand and compensate for the fatigue in class with a little-and-often approach and to plan to compensate for the deconditioning when returning to class after a period of rest such as a holiday.

Psychosocial markers of JH that are also relevant to dance

Whilst it is established that JH is prevalent in dance because of its associated physical or biological characteristics (Armstrong & Greig, 2018; Chan, Hopper, Zhang, Pacey, & Nicholson, 2018; McCormack et al., 2004), there are also psychosocial aspects of JH that make it potentially less conducive to dance training and performance. These are again things that dance teachers should be aware of, understand and work towards mitigating . For example, JH has more recently been unexpectedly associated with increased anxiety (Bulbena-Cabré et al., 2018). This has since been further investigated from a psychosocial perspective and JH is increasingly becoming also connected with poor body awareness and disorders that include anxiety, fear and dysautonomia (a condition in which the autonomic nervous system (ANS) does not work properly and adversely affects the health and may affect the functioning of homeostasis, the heart, bladder, intestines, sweat glands, pupils, and blood vessels) (Baeza Velasco et al., 2016; Bulbena-Cabré et al., 2018). Unlike the biological characteristics of JH, these are clearly less conducive to a training in dance and it is therefore important for teachers to know and recognise the signs and implications of these conditions which are further explained below.

Anxiety and bodily crisis in JH

Within 'normal' functioning and a balanced body awareness there is a threshold, for example to anxiety or window of tolerance, within which we all operate. Trauma or turbulence, potentially caused for example by the symptoms and conditions that present within JH (such as poor or imbalanced body awareness and/or additional stressors such as a feeling of being overwhelmed and anxious), can potentially result in a person with JH 'flipping' between and or getting temporarily stuck within either the hyper and hypo arousal states (Ogden, Minton, & Pain, 2006; Siegel, 2010). This is further explained using the polyvagal theory and the notion of our three defence mechanisms (fight, flight and dissociation) that are triggered when danger or turbulence is sensed (Porges, 2017, p. 101).

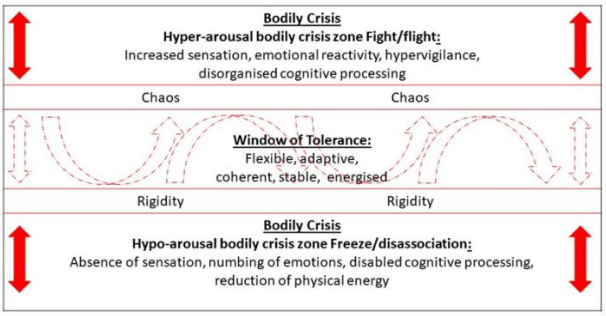

Figure 1 A model of Bodily Crisis within JH adapted by Timmons (2020) and applied to Polyvagal and Window of Tolerance theories. Adapted from Siegel and Ogden, Minton & Pain (Siegel, 2010; Ogden, Minton, & Pain, 2006)

Creativity, emotional expression, learning and JH

Interestingly, the polyvagal theory further proposes that when the body is in any one of these defence mechanisms it is using metabolic resources and cannot for example be creative or heal (Heilman et al., 2008; Kemp, 2017; Porges, 2017). The implications for this within dance are significant as creativity is fundamental to the art form and injury is prevalent within dance due to the demands at a professional or vocational level. Porges (2017) further proposes that the "neural pathway for healing overlaps with the neural pathway for social engagement" (p 101) and it is indeed the vagal pathway that sends information between the brain and the body signalling safety and providing calm. This pathway, in addition to providing feelings of safety and calm, also enables social engagement and expression in the face and body (Porges, 2017, p.101-102). Again this is significant in dance as expression through the face and body is critical. Therefore the neural pathway for social engagement that Porges discusses, including the interchange of brain-body signalling, can potentially also include behaviours that are pertinent to effective teaching and learning environments. For example, 'feeling safe' is a condition that is conducive to learning as a form of social engagement. Indeed a recent paradigm shift within learning theory and practice now recognises the important role of emotional well-being for learning alongside the more established theories of motivation (Rowe, Fitness, & Wood, 2015). It is now understood that intense emotions are associated with hypermobility (Eccles et al., 2015), and intense emotions can also invigorate the homeostatic mechanism and influence the vagal states (fight, flight and disassociation) within JH. Intense emotions are also however a catalyst for art and in such "fuel the dance-making process and ignite audiences'' (Minton & Faber, 2016). Indeed good dance training involves the cultivation, manipulation and communication of emotions from a relatively young age. This then suggests that appropriate teaching awareness and understanding of these mechanisms within the dance environment may act as a protective factor for dancers with JH as it will inevitably enable a deeper understanding of feelings and emotions and hence unavoidably also self-regulation.

For these reasons it is important that all teachers of dance develop their awareness of and understand the bio-psycho-social implications of JH as it is unavoidably prevalent in dance settings. For teachers of dance in inclusive settings it is further important that they understand not only the complexities of working in an inclusive environment but also the potential interrelatedness of, for example, DS, ASD and JH.

This section is based on research included in the PhD thesis submitted by Wendy Timmons who is programme director for the MSc Dance Science & Education at the University of Edinburgh.

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