

# **Text Summary**

# Juvenile Hypermobility and CRPS With Dr. Nathan Hasson

About Dr. Nathan Hasson

- Nathan has been a medical practitioner for many years.
- Originally from Zimbabwe, he was practicing in Cape Town 30 years ago before coming to the UK.
  - Moved to the UK at this time to allow wife to do her Master's degree.
  - Dr. Hasson was able to meet the people that 'wrote the books'.
- Began working at the Hammersmith Hospital, looking after children with muscle
- diseases.
  - Previously working in pediatrics, it was here that Dr. Hasson learned how to assess
  - muscle strength in children.
  - Here, he mainly worked with boys who suffered with muscular dystrophy and children with neuromuscular disorders.
- Then, he moved on to Northwick Park Hospital to extend his practice into the world of rheumatology.
  - Good mix of specialism to be a true 'musculoskeletal person'.
    - Became a part of the first group of musculoskeletal pediatricians with
    - knowledge of both neuromuscular and rheumatological disorders.
    - E.g. Overlap when children had dermatomyositis.
- Learning the skill of examining joints was important.
- He lectures to all sorts of people across the UK.
- Maintains regular contact with multi-disciplinary practices including Steven Sandler in the osteopathic world.

Patient range and type

- Whilst working in pediatrics, Dr. Hasson would see patients ranging between 4-18.
  - (You can't do muscle strength under the age of four, appropriately, using manual muscle testing.)
- Although rare, he looks after children with juvenile arthritis.
- More common is issues related to musculoskeletal problems.
  - Today, children are actually very weak physically.

- E.g. Children who are hypermobile do not experience problems because of this, but rather that they are weak physically.
- An Australian study found that modern ten year-olds were a minute and a half slower than their parents were over a mile at the same age.
  - This equates to more than a lap slower around a 400m track.
- This is becoming a worldwide phenomenon through lack of physical exercise.
- Other interesting ties include low Vitamin D levels and obesity.
  - In fact, children consume 2,500 calories a day in comparison to their parents' generation who ate 3,330 calories a day.
  - The 30% difference in obesity levels is caused by a lack of activity.

## Use of the term Reflex Sympathetic Dystrophy (as opposed to CPRS)

- It's better for something's name to describe what it is.
  - E.g. Osgood-Schlatter, or any named conditions, does not say what the problem is.
- Similarly, Complex Regional Pain Syndrome is not an effective conveyor of the issue.
- Conversely, Reflex Sympathetic Dystrophy explains the condition well.
  - The reflex is allowed by the body, but not it's not something that was planned.
  - It's the sympathetic nervous system which mediates the pain- therefore pain killers will be ineffective.
  - Dystrophy is effectively a final consequence when there is lack of usage.
- These three words explain exactly what is going on and can be used to enlighten parents.

# Joint Hypermobility Syndrome (Benign/Differences etc)

- Named by 'Mother of Pediatric Rheumatology', Barbara Ansell, and a colleague in 1967.
  - Found that sometimes people develop musculoskeletal aches and pains not related to arthritis or some other rheumatic condition.
    - She recognized that sometimes people who are flexible develop aches and pains.
    - At the time, they did not realize that the association with flexibility and pain is more to do with being physically weak.
- Refined to Benign Joint Hypermobility Syndrome in children.
  - Children don't suffer damage from being flexible and weak- they can improve.
- Used to be called 'growing pains'.
  - In actual fact, it is joint hypermobility syndrome whereby the relative lack of muscle strength for the new person, compared to the 'old' person causes aches and pains.
- Rheumatologists do not tend to use the name, 'Juvenile Hypermobility Syndrome'.

# Case History: Jake Newitt

(This part of the dialogue contains comments from J: Jake (patient), B: Bob (parent) as well as Dr. Hasson.)

- J: Experience with JHS was 'incredible'- really painful, yet there is nothing that you can do.
- It can begin in one part of the body and then spread into multiple areas.
- Believe to have begun after a car crash but did not fully develop until a little later.
  - Could also be attributed to a footballing incident.
- Reduced to a wheelchair when mild pain increased.

- N: Reflex Sympathetic Dystrophy is often triggered by an event.
  - This is why the word 'reflex' is so vital as it is developed by the body to protect.
    This leads to the massive cascade of pain.
- The medical word is allodynia- pain that is being caused by something that can't cause you pain.
- Like with Jake, reflex sympathetic dystrophy can flit between different areas of the body.
- Standard pain could be lessened with the use of painkillers.
  - Pain from Reflex Sympathetic Dystrophy is mediated by the sympathetic nervous system and as such, morphine and others would have little effect.
  - Distraction has a greater effect as it temporarily switches off the sympathetic nervous system.
    - A lack of answers limits the amount that one can be distracted.
- B: Forced to have time off work due to seven straight weeks of severe pain before seeing Nathan.
- Getting knowledge from Nathan was vital for hope that things could be turned around.
- N: People that experience RSD also tend to be 'worriers'.
- Teaching people to relax can help distract them from the pain.
  - One thing to note is that RSD is a 'useless pain'- this can be used to help the patient to relax with the pain.
- Immediately with Jake, he was made to get out of his wheelchair and it on the bedalmost confronting the pain.
- J: Hated the experience but in hindsight, it was the first step towards recovering.

#### State of GPs' Knowledge Towards RSD

- B: The GP was googling symptoms during the appointment.
- Possibly more knowledgeable than a GP thanks to Nathan's explanations.
- N: Every medical practitioner has their own set of skills.
  - If you've never come across this type of condition, you would not recognize it.
  - People also do not understand that painkillers have no effect.

#### Seeing Nathan

- N: Jake was very weak upon first examination.
  - Understandable as he was unable to do anything.
  - He had elbow and leg pain as well as backache.
- Although difficult for Jake to get out of his wheelchair and on to the bed, it had to be done.
- It is a possibility that if they are not pushed, they could permanently end up in a wheelchair.

- Touching and desensitizing the areas of pain may sound like they are doing harm but in fact, they're (as a parent) getting the child's nerves to understand that it is normal.
- It is draining to treat a child with reflex sympathetic dystrophy because you suffer with them- but there isn't another way.
- The same desensitizing exercises are recommended to parents and children.
  - As above, this determines and maintains that the area is safe to be touched.

#### The Process

- B: Jake experienced six months of pain before seeing Nathan.
- This was after numerous trips to the GP and A&E.
  - Hard to know as a parent what to do.
- The GP or Hospital would administer painkillers and the bursts of pain would naturally subside temporarily, before beginning again.
- N: Unfortunately, some people believe the disease is made up and that it is just a psychological thing- once they understand it, it seems obvious.
- J: First few months of treatment are extremely difficult
  - Important that his dad pushed him to keep up exercises and carrying out daily actions (like eating dinner without help- which had become impossible due to
  - pain.)
- N: Complete exercises with somebody else- more motivating together rather than exercising whilst watching the rest of the family watch TV.
- J: Eventually, the exercises become much more bearable and finally, almost easy.
  - Carried them out twice a day.
    - Despite feeling much better now, continued exercise is essential to avoid a relapse into renewed pain.
- B: The child has to do the exercises themselves.
  - If they carry out the exercises, change can be dramatic in a matter of months.
- N: They're given a program of muscle-specific exercises which start off with just five repetitions.
  - Building up: quads, around the ankle etc.
  - Increasing the number of repetitions/ weight (around the ankle) over time.
- Hydrotherapy is, however, the opposite of what is needed.
  - It causes the muscles to relax, not to build up strength and stamina.
  - For this reason, it is great for conditions like Arthritis.
  - Dry land exercises are much better for rebuilding.
- N: Certain tools such as 'Buzzy' (which was actually invented as a device to distract children having blood tests in America) can be useful desensitizing agents.
  - The vibration of the tool is extremely painful originally, however it becomes bearable and actually has a positive effect when the skin is touched naturally.
  - Similar to a TENS machine- but children don't like being electrocuted!
- The tool is particularly useful as it encourages the patient to move.
  - The desensitization of the body part makes them feel more comfortable.

• Movement is vital in the process.

#### School

- J: School have been very good.
  - Happy to help and support if anything happened.
- N: Distractions are very useful- for this reason, returning to school is helpful.

#### Multidisciplinary Team

- N: Best team to recommend to patients is at Great Ormond Street.
- Sue Maylard and the team of psychologists, physiotherapists and OTS do not baulk at this.
  - They know that if you stop desensitizing an area, the patient will not get better.
- B: Initially, it was possible to see Sue.
- Then, due to factors such as catchment areas, Nuffield in Oxford was an easier option.
- From knowledge gained from practitioners, it was possible to 'be the boss' at home.
- Simply necessary to shut out the agony of the pain and continue to desensitize.

#### Pain Management Groups

- N: Must be careful with groups.
  - Statistically, some groups can actually have negative effects on a patient.
  - Naturally, other groups can be extremely positive.
    - Help needs to come from the right direction and you need the correct advice.
- B: The support group at Oxford is made up of a large age range of children- 5-17 years old.
- Everybody has a chance to explain their situation.
  - Jake gave a lot of hope to the group.
- Interestingly, Jake's knowledge was staggeringly better than many of the other children.

#### Dietary matters

- N: Vitamins don't have anything to do with this.
  - Lack of Vitamin D is a problem in society but it is unrelated to CRPS.
  - Only tenuous link is that the deficiency is caused by lack of activity outdoors (similar to children's weaknesses).

#### Prognosis

- N: If you build up strength and stamina and understand CRPS, you do not let it come back.
- Certain people who suffer with it will see it move around to different parts of the body.
- Important to get your psyche route not listening to unimportant pain.

#### In Clinic

- Expect to see somebody who has been in pain, however they may not be at the time.
- They have not responded to analgesics.

- Look for hypermobility in the patient (double-jointed etc.)
- The Beighton Criteria is a nine-point scale of measuring hypermobility.
  - However, the majority of points relate to the upper-limbs (many children seen by Nathan often have lower-limb complaints)
  - In addition, the only lower-limb test within the Beighton Criteria revolves around the hyperextension of the knee which is no longer the norm in children.
- The Bulbena Criteria looks at hip movements, flat-footedness, patellar instability as well as how easily somebody bruises.
- More extensive than the Beighton.
- You can find out about the whole ten-point criteria online- how to examine etc.
- Manual testing can compare the strength of muscles and obviously determine weak areas.
  - There are 10 muscles measured in the upper-limbs, five on each side. Children today only have normal right biceps.
    - Possible to overcome the shoulder abductors of teenagers with one finger but I can't overcome their mothers'.
  - There are 12 muscles tested in the lower-limbs: the hip flexors, the hip extensors, the hip abductors, the hamstrings, the calves, tibialis anterior.

Only hamstrings and calves are considered normal in the lower-limbs.

- In total, five out of twenty-two muscles tested are considered normal.
- The Oxford Scale of five is used to grade strength in manual muscle testing:
  - Five is normal strength
    - Four is strength against some resistance but it can be overcome.
    - Three is antigravity muscles.
      - Some childrens' inner-range quads (vastus medialis) are weaker than gravity.
      - Can lead to chondromalacia patella (common now: 1 in 3 teenagers)
- It is becoming so that you can tell the difference between a normal and a person who has a disease through 1 or 2 muscles.
  - Before, a boy with (e.g.) Becker muscular dystrophy had 10/22 normal muscles and everybody would have 22.

#### **State of Children generally**

- Children nowadays are physically weak.
  - At age four, children's muscle strength is much weaker than 28 years ago.
  - Rebuilding is essential but must be done in a fun way to engage child.
- Strength and stamina is essential if you are a flexible person.
- When confronted with conditions such as; CRPS, hypermobility and fibromyalgia, anxiety is often the biggest enemy.
  - Negative thoughts about movements and actions lead to suffering and deterioration.

#### Adult Hypermobility

- First difference is that the condition is benign in children.
  - Children don't suffer the damage that happens in adults with hypermobility.
  - They do not slip discs, wearing and tearing joints or getting osteoarthritis.
  - Children can develop CRPS, tiredness, aches and pain.
- Many hypermobile adults have not experienced problems during childhood as they had decent strength.
  - If you were a gymnast when you were young but then stopped in busy adult life, that's when problems develop.
  - In particular, when women have babies, discs L4-L5, L5-S1 start to go, neck discs start to go, shoulders start to freeze.
    - Many people who have flexibility and disc problems will tell you as soon as they start doing Pilates, as soon as they start doing swimming, as soon as they start rebuilding their strength and stamina, they find their pain goes away.
  - If you stop doing exercise, then you are going to end up having problems related to your flexibility.
- No difference in prevalence between children and adults: if you're born flexible, you're flexible.
  - Dependent on population group:
    - In South American countries, about 40% of people are hypermobile.
    - Not a gene defect, rather a normal variation.

Sports

- Yoga is not as good for hypermobile people as it is fairly static
  - Essentially, they find it easy
- Conversely, Pilates physically builds strength and stamina, as well as core muscles.
  - This protects the joints and improves pain in adults.
- In effect, hypermobile people are an enhancement of a normal person:
  - It's impossible to make yourself hypermobile.
  - If you're strong, flexibility can help you become the best sportsperson but weakness can cause trouble.
- No sports to avoid if you are hypermobile- the exercise will help you to avoid trouble.
- If you're strong enough, you end up in a vicious cycle of aches and pains, of tiredness and then as an adult with damage and it's hard to come back from damage.
- Becoming physically fit and strong then means you will have more energy and be able to keep going longer in the day.

### **Ehlers-Danlos**

- Flexibility all comes under the name hypermobile.
  - All have flexibility in the joints.
- Dr. Hasson therefore does not like to use the classes of Ehlers-Danlos to differentiate between both flexible skin and joints and just flexible joints.
- Ehlers-Danlos type 4 is an exception.
  - It is a known genetic problem which can result in burst blood vessels.
  - There's no treatment for this and it is unfortunately fatal.

• The rest of the 'types' would all have similar specific muscle exercise programs.

#### Is taping useful at all?

- Strapping and taping does appear to stimulate the underlying muscles to work better.
  - E.g. if one is more prone to developing scoliosis.
  - Doing physio and strengthening muscles is the best solution.
- Kinesio tape probably would not distract somebody who has developed RSD.
  - However, if somebody is weak and flexible, it could be useful to prevent and twist (etc.) which could trigger RSD.

#### Tubigrip

- SB: Was its purpose not so much as a joint supporter but as a stimulus to get your brain thinking it was playing that role?
- N: However, such an object can also become a crutch.
- Practitioners can be quite quick to put somebody in a splint rather than build-up their muscles.

#### Areas affected in people with hypermobility

- Joints
- Skin
  - Bruising easily is a consequence of this and is on the Bulbena Criteria.
  - Some parents have had run-ins with Social Services but it is purely down to
  - flexibility.
- Possibility of getting gastroesophageal reflux
  - The gastrointestinal tract is affected.
  - The lower esophageal sphincter is very law
    - The babies suffer from gastroesophageal reflux.
    - As adults, they can have heartburn and indigestion.
- Hypermobile people sometimes have lower blood pressure.
  - Less chance of heart problems.

However, if you don't have a good muscle pump in your quads and you stand up and you don't pump blood up, your blood pressure drops and then you get like a moment of dizziness or a head rush.