HYPOTONIA

An Overused Term

Not a Diagnosis but a Symptom ???

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First Early Signs

What Is It?

- **The amount of tension or resistance in a muscle to passive stretch – not the same as strength or joint mobility**
- **Varying degrees of involvement**
- **Abstract, Imprecise and Undefined – subjective term, no objective way to measure -which leads to lack of knowledge of the underlying mechanism** –

Martin,,K, Innman, J. et. al, " Characteristics of Hypotonia: A Consensus Opinion of Ped. OT and PT" *Pediatric PT*, 2005

Typical Criteria used for Early Dx. of Hypotonia

- Head lag on pull to sit
- " Slipping thru" when held under shoulders
- Increased Open Angles at Joints including popliteal angle & scarf sign
- Not bearing weight on legs
- Gross Motor Delays

Mintz- Itkin, R, Lerman-Sagie, T, et.al ., " Does PT Improve Outcome in Infants with Joint Hypermobility and Benign Hypotonia?" *Journal of Child Neuro*. 2009

Hypotonia

- Inconsistency for identification, referral and treatment
- Few studies done investigating children with hypotonia when no underlying cause is found. This group remains a challenging dx for many physicians and a frustrating one for parents of infants given the dx.

Strubhar,A,Merand,K,Morgan, A."Outcomes of Infants with Idiopathic Hypotonia" *Pediatric Physical Therapy*, 2007
Martin,,K, Innman, J. et. al, " Characteristics of Hypotonia: A Consensus Opinion of Ped. OT and PT" *Pediatric PT*, 2005

Hypotonia Associated with Many Different Conditions

- Genetic Disorders (most common)
- Central Nervous System Disorder
- Motor Unit
- Autoimmune disorder
- Infections
- Metabolic
- **Toxins**

Stubhar,A. , Meranda, K , Morgan, A " Outcomes of Infants with Idiopathic Hypotonia"Pediatric Physical Therapy, 2007
Martin,,K, Innman, J. et. al, " Characteristics of Hypotonia: A Consensus Opinion of Ped. OT and PT" *Pediatric PT*, 2005

Important to Try and Find The Underlying Disorder

- Detailed and thorough family, pregnancy, birth and early motor development history
- Early Onset? Or later? Has there been improvement over time or progression of symptoms ?
- Referral to Neurologist, Genetics and/or Developmental Pediatrician or specialist most experienced in diagnosing muscle disorders

Thompson,CE, "Benign congenital hypotonia is not a diagnosis". *Dev Med. Child Neurol.* 2002

Does " idiopathic" hypotonia resolve as child gets older ?

- Some research states that this type of hypotonia resolves within the first few years in majority of children
- Other studies have found mild deficits remain including clumsy gait and speech delays (followed till age 4)
- Other studies found no significant difference in these children later on (6 to 8 years) except in gross motor performance
- Study following children to 12 years based on parental and teacher questionnaire found dx of hypotonia was related to problems in gross motor and behavior

Strubhar, Meranda, " Outcomes of Infants with Idiopathic Hypotonia" *Ped. PT* 2007
Mintz-Itkin, r. Lerman- sagie, et-al " Does PT Improve Outcomes in Infants"..*J Child Neurol,* 2009

IF No Clear Etiology and Isolated Finding ????

In past often referred to as :

- Benign Congenital Hypotonia – old term - Walton-1957
- Congenital Hypotonia with favorable outcome
- Idiopathic Hypotonia
- Perhaps the most poorly understood group.

Much disagreement in literature of what to call this condition and if it really is benign or even if it exists in isolation of other findings.

- True incidence is not known but terms less commonly used due to underlying dx. being found.

All other causes should be ruled out by thorough work up.

Stubhar,A. , Meranda, K , Morgan, A " Outcomes of Infants with Idiopathic Hypotonia"Pediatric Physical Therapy, 2007
Mintz- Itkin, R, Lerman-Sage, T, et.al ., " Does PT Improve Outcome in Infants wit joint hypermobility And Benign Hypotonia? Journal of Child Neuro. 2009

Outcomes- Small Study

- 105 children dxed with hypotonia in infancy

average rate of dx was 11 mos families contacted three years to 13 yrs plus median age was 8yrs,

Four groups –

10% - no other dx made

32% - l.d., language delays or ADHD

40% - M.R, Developmental or Genetic Disorder

16% - other Dx. including CP

Stubhar,A. , Meranda, K , Morgan, A " Outcomes of Infants with Idiopathic Hypotonia" *Pediatric Physical Therapy,* 2007

When Isolated Finding ??

- Evidence also exist that this group with no known underlying cause and isolated finding of having an increased incidence of family history of delayed motor skills in 30% of the cases.
- Varied reports and few studies done on future outcomes of these children or degree of residual problems past age 3

Mintz- Itkin, R, Lerman-Sage, T, et.al ., " Does PT Improve Outcome in Infants wit joint hypermobility And Benign Hypotonia? Journal of Child Neuro. 2009
Stubhar,A. , Meranda, K , Morgan, A " Outcomes of Infants with Idiopathic Hypotonia"Pediatric Physical Therapy, 2007

PT and OT Consensus

- Decreased Strength
- Decreased activity tolerance
- Decreased motor skill development
- Rounded Shoulder Posture
- Hypermobile Joints
- Increased Flexion in Joints
- Poor Attention and Motivation

Stubhar,A. , Meranda, K , Morgan, A " Outcomes of Infants with Idiopathic Hypotonia". *Pediatric Physical Therapy,* 2007. Martin,,K, Innman, J. et. al, " Characteristics of Hypotonia: A Consensus Opinion of Ped. OT and PT" *Pediatric PT*, 2005

What Functions ?

- Gross motor skills- motor control
- Fine motor skills - visual motor
- Oral motor or suck, swallowing skills
- Expressive language
- Sensorimotor skills
- Social ??? Poor Motivation to Move Research varies and still questions about Poor Attention ? Learning ? Cognition ?

Martin,,K, Innman, J. et. al, " Characteristics of Hypotonia: A Consensus Opinion of Ped. OT and PT" *Pediatric PT*, 2005

Delayed Milestones and Poor Quality of Movement

- Just sits, scoots or rolls to move >9 mos
- Delayed pull to stand and walking
- Keeps wide base- frogged leg position
- Poor weight shifting , falls often, tires easily
- Overall increased joint mobility with pronated - everted feet
- Later - poor push off for running and higher level skills such as galloping and hopping
- Poor balance on one foot
- Poor posture – sitting and standing

Gross Motor Skills

- Poor quality or graded control
- Transitions of movement
- Motor Control or Motor learning
- Early postural changes
- "Hanging on joints"
- Child develops compensations
- Head back
- Wide based arms and legs ' frogged"
- Moves with less weight shifting or trunk rotation
- Decreased Joint Proprioception

What To Do?

- Early PT , OT , ST and cognitive stimulation
- Decrease the compensations
- Increase the motor control and ability to transition with more "normal" movement patterns
- Practice and carry over through out the day, through handling, positioning and home programs beginning very early

Hypotonia 3 to 6 mos

- Poor head control - not holding head well by 3 months, or stacking, not righting head by 4 months when tilted in supported sit, not bringing hands to midline or asymmetrical
- Poor trunk control - rounded back at 6 months, not lifting head well in prone or reaching . Not righting trunk or head with weight shifting in prone or in space , not sitting by 6 to 8 mos

Physical and Occupational Therapy

- Improve Strength
- Head, shoulder and trunk control then leg strength
- Improve Posture in all positions
- Intervene before the compensations develop
- Strengthening and Balance programs with parent involved
- Good seating posture in high chair , toddler seat, school chair
- Standing with help by 10 to 12 months

Physical Therapy continued

- **Practice Standing ? Standers ?**
- **Orthotics-pre-fab, or custom – improve alignment of foot/ankle**
- **Adaptive Walkers – age 2 +**
- **Hip Helpers**
- **Adaptive tricycles / pedals with straps**
- **Small group classes Gymboree's, etc**

Strength Training

- Children with atypical motor development have limited repertoire of movements and sometimes paucity of movement
- Compensatory Movement strategies learned in early development lead to decreased strength and endurance of common key muscle groups

Bundonis, J. "Pediatric Strength Training " *Rehabpub.com* 2007

Does PT and OT help babies with hypotonia ???

- No agreed treatment approach, frequency, and protocols
- Based on professional tradition and not evidenced based
- Few reliable guidelines for intervention most often NDT / Bobath approach used for babies
- Schreiber found an inverse ratio between the parent ability to exercise with baby and the treatment session frequency

Mintz-Itkin, r. Lerman- Sagie, et-al " Does PT Improve Outcomes in Infants".*J Child Neurol,* 2009

Strength Training

- Focus on thoracic extensors – often an overuse of pectorals – rounded shoulders
- Abdominals – core strength (flared rib cage, difficulty sitting up straight , difficulty flexing against gravity
- Hip Extensors – maintain wide base
- Quadriceps – especially terminal knee extension

Bundonis, J. " Pediatric Strength Training" *Rehabpub.com* 4/2007

Small Study

29 infants – two groups (8 to 12 mos) dxed with idiopathic hypotonia

NDT approach & parent home instruction

- Monthly session
- 9% walked at 15 mos
- Weekly session
- 45% Walked at 15 mos = WNL

Early benefit did not continue at 18 mos.
100 % were walking in monthly group and all but 2 in weekly group
No Difference in AIMS Score on Quality of Movement
AIMS scores were still smaller or equal to the fifth percentile

Mintz-Itkin, R. Lerman- Sagie, et-al " Does PT Improve Outcomes in Infants". *J Child Neurol.* 2009

Core Strengthening

Hypermobility – Hypotonia

- Often used interchangeably
- Really two separate entities

Hypermobility vs. Low Muscle Tone – Two Separate Entities

- Many children with low muscle tone also have increased joint mobility and decreased strength and literature showed correlation

However:

- Not all children with hypermobility have low muscle tone or decreased strength
- Joint hyper mobility common in childhood , Prevalence depends on age, sex, ethnicity and decreases as child gets older.
- Hypermobility and hypotonia often frequent findings in infants with delayed motor milestones but are not the same entity

Mintz-Itkin, R. Lerman- Sagie, et-al " Does PT Improve Outcomes in Infants". *J Child Neurol,* 2009

Joint Hypermobility

- A condition that features joints that easily move beyond the normal range
- Normal to have increased joint mobility in infants and young children and joint mobility decreases with age
- Can be isolated finding of only one to a few joints or multiple joints, or an aspect of a clinical syndrome

Mintz-Itkin, R. Lerman- Sagie, et-al " Does PT Improve Outcomes in Infants". *J Child Neurol,* 2009
Murray, K " Hypermobility disorders in children and adolescents " *Best Practice & Research Clinical Rheumatology* 2006

Research- Orthotics

- Studies have shown orthotics helpful to increase stability for children with low muscle tone
- Martin looked at postural stability of 17 children with Down Syndrome with flexible SMO to decrease pronation showed significant improvement in Gross Motor Function Measure (GMFM) and Balance subtest of Bruininks – Oseretsky Test of Motor Performance (BOTMP) for increased stability for walking, running, and jumping after 7 weeks of wearing SMO's compared to wearing only shoes

Martin,K. Effects of SMO on postural stability in children with Down Syndrome Neurol. 2004;46:406-11

Joint Hypermobility Incidence

- More common in girls than boys and some ethnic groups, runs in families
- Prevalence ranges from 6.7% to 39.6% depending on population and criteria used
- Usually gets better or remains stable with age
- Hypermobility of ankle , hip and elbow joints was found to have best correlation with gross motor delay

Mintz-Itkin, R. Lerman- Sagie, et-al " Does PT Improve Outcomes in Infants". *J Child Neurol,* 2009

QuickTime™ and a
decompressor
are needed to see this picture.

Orthotics

- Can help many children
- Low tone or child with increased joint mobility , including pronated feet when complaints of tiredness and pain in legs or knees when walking long distances -
- Toe walkers – compensation
- Types vary from pre fab arch supports to custom molded, UCBL's , SMO's
- Children who have more impairment and need control above ankle = AF0's hinged or solid

Joint Hypermobility

- Common finding in other connective tissue disorders including Marfan syndrome, Ehrlos Danlos and Osteogenic Imperfecta.
- Often finding in patients with Juvenile Rheumatoid Arthritis and Chronic Pain Syndromes
- Dx finding similar with low tone, genetic and metabolic disorders

Everman DB, Robin NJ. Hypermobility Syndrome Pediatric Rev. 1998 19:111-117

ORTHOTIC CHOICES

- None – supportive but flexible sneaker/ shoe
- Pre Fab
- UCBL
- Custom Molded
- SMO –
- AF0 – hinged or solid
- PLSO

Joint Hypermobility Syndrome

- Remains unclear why some children who have generalized hypermobilty become symptomatic as they get older with musculoskeletal complications while others remain symptom free
- Fatigue, Widespread Muscular Skeletal Pain , Joint Pain and Instability Episodes with no evidence of any rheumatic, neurological, skeletal or metabolic disorder
- Soft Tissue Injuries , Osteopenia
- De conditioning

Tofts, l, Elliiot et. al. " The differential diagnosis of children with joint hypermoblilty : A Review of the literature" *Pediatric rheumatology online journal* 2009

Joint Hypermobility Syndrome

- As in low tone, joint hypermobility more common in African, Asian and Middle Eastern descent
- 5% of females have JHS compared with .6% men
- Strong genetic component with an autosomal dominant pattern with first degree relatives with the disorder can be identified in approx. 50% of cases

Seclin, U, Sonel etal. " The prevalence of Joint hypermobility among high school students"*Rhematology Int* 2005;25;260-263
EngelbertR, Uiterwaal C, et al Pediatric generalized jt hypomoblity and musculosckeltal complaints: a new entity? *Pediatrics* 204;113:714-719

Joint Hypermobility Syndrome

- Appears to be an abnormality in the collagen subtypes
- Mutations in the fibrillin gene have also been identified in families with JHS

Magnusson Sp. Viscoelastic properties and flexibility in benign joint hypermobility Syndrome *J Rheumatol.* 2011;28:2720-2725

Formulate a Management Plan with use of

Interdisciplinary Team investigate if underlyin disorder

and management of symptoms

Tofts, I, Elliiot et. al. " The differential diagnosis of children with joint hypermoblilty : A Review of the literature" *Pediatric rheumatology online journal* 2009

Physical and Occupational Therapy for Children with Joint Hypermobility Syndrome

- Improve strength to increase joint stability
- Assess all joints for compensatory tightness
- Improve joint proprioception & balance
- Improve endurance for gross and fine motor activities and learning to " pace" activities
- Modifying activities to low impact if needed
- Assist with sitting and standing posture
- Assist with fine motor strength, handwriting
- Decrease pain by becoming more strong and fit

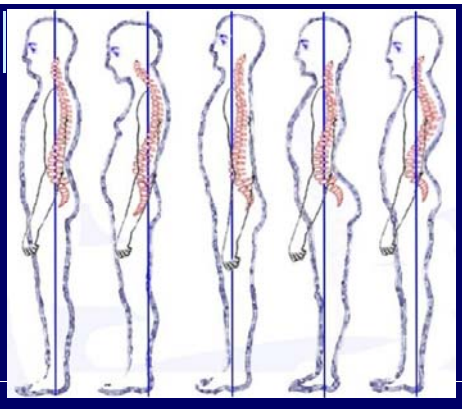
Fatoye,F,Palmer,S. "Proprioception and muscle torque deficits in children with hypermobility syndrome" *Rheumatology*,2009
Murray, K " Hypermobility disorders in children and adolescents " *Best Practice & Research Clinical Rheumatology*, 2006

?????? During History

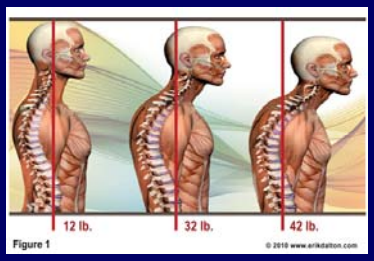
Patient Interview Questions to Ask

- Can you now (or could you ever) place your hands flat on the floor without bending your knees?
- Can you now (or could you ever) bend your thumb to touch your forearm?
- As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
- As a child or teenager, did your shoulder or kneecap dislocate on more than one occasion?
- Do you consider yourself double-jointed?

Simpson, Michael, DO *JAOA Clinical Practice* ; "Benign Joint Hypermobility Syndrome : Evaluation, Diagnosis and Management " Vol106, No.9 9/2006-531-536



Back Packs

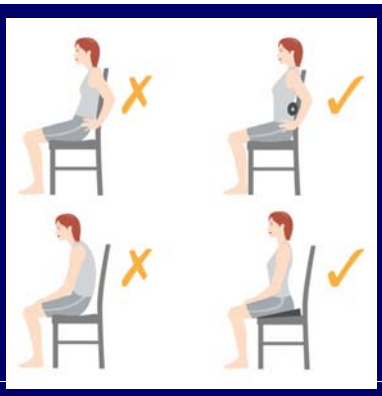


Children should not carry more than 10 to 15% of their body weight and less is better

http://orleansgardenschiro.com

Foot Pronation vs. Flat Feet

- 2 degrees normal and necessary
  Often heredity , low tone or spastic foot
- Can lead to c/o of knee pain, tiredness when walking
- Tight heel cords
- Navicular pain



Calcaneal Valgum

When To Wear Orthotics?

Supportive Sneakers -Shoes

- Boots, high tops, sandals, flip flops , clogs not advised for daily wear
- Good supportive shoe that fits well can often make a big difference.
- Need stability at heel counter, flexibility in toe box

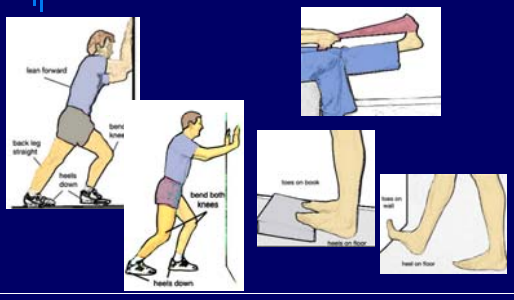
- Mid foot collapsed caused by tight heel cords which leads to foot pronation and then mid foot collapses = painful foot when walking

Common with spastic foot

Also happens with low tone weak foot mm from early excessive foot pronation which leads to tight heel cord and can then produced mid foot collapse and painful foot

- **The normal motion of the leg moving forward over the foot is shown here. If the calf muscles are tight and do not allow this movement at the ankle, the foot must flex instead of the ankle.**
- **The result is flexion (and collapse) of the midfoot, and movement of the front of the foot out (away from the midline of the body) to allow the leg to move over the foot and then propel the body forward. In the image above, the motion is simplified.**

Heel / Foot Stretching



Activities - What to recommend?

- Low risk injury
- Walking,running, biking, hiking, swimming, T-ball, yoga, tai chi,
- Karate,golf, bowling
- * Avoid jumping from high surfaces - passive neck flexion/extension
- Higher risk [falls]
- Horse back riding
- Rollerblading/ice skating
- Skiing
- Soccer - no heading
- Light weights

Tips for Success for Exercise

- Everyone can always work on strength, endurance and coordination - life long task
- Choose something child/ adult likes
- Build in fun and variation
- Keep the competition down
- Make it happen - several times a week
- Always wear appropriate protective gear