

To Find out the Value of Hypermobility in Down's Syndrome

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ABSTRACT

Background: Down syndrome, the result of trisomy of chromosome 21, is one of the most common chromosomal abnormalities. Patients have a characteristic 8 facial appearance, variable levels of intelligence and self-care skills, and a variety of associated medical conditions. Orthopaedic manifestations occur frequently; most are related to hypotonia, joint hypermobility, and ligamentous laxity.

Objective: Aim of the study is to find out the value of hyper mobility in down's syndrome.

Methodology: 30 Subjects including in my study ,study duration is 4 weeks, study setting is Maithree Special School, NIEPMD, REC Centre SRM Hospital.

Outcome Measures: Beighton Scale, Goniometre

Results: This beighton scale consists of goniometer measurement of thumb, metacarpophalangeal joints ,elbow joint , knee joints, trunk flexion .total point (9) point.50%of sample was scored 9 point.10%of sample was scored 8 point.7%of sample was scored 7 points.35%of sample was scored 6 points.

Conclusion: This study concluded that Down's Syndrome children has hypermobility which needs to be investigated early and treated for better quality of life.

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Introduction

Down's syndrome is the most commonly identified genetic chromosomal anomaly world-wide. For past 100 years the identification of Down's syndrome was done according to the clinical significances. Down syndrome has been estimated to occur ~1 in 732 infants in the United States. And the prevalence of DS in India (Mumbai) is 14.2%. Down Syndrome characterized the particular type of genetic errors due to missegregation (i.e.) late or instable pregnancy and birth defects [1].

Approximately 27% of the individual had lax joints. Men's joint laxity declined with age although in mid –twenties, women joint laxity continued through the mid forties [2]. More than 6000 children were evaluated for hyper mobility with Beighton score 13.8 years was the average age group of the study and percentage of girls and boys was 27.5% and 10.6%. Greater than 4 score in Beighton score was too low and appropriate for the children in UK for musculoskeletal development [3].

Hyper mobility is that stretch further than normal, it is otherwise called Benign joint hyper mobility. Syndrome (BJHS), hyper mobility syndrome (HMS).Ehlers-Danlos Syndrome (EDS) are

genetic connective tissue disorders, symptoms like loose joints, joint pain, abnormal scar formation. Laxity of joints and muscles are interrelated with each other which also leads to change in the ROM various individual with age, race and body build. In both sex and age related, racial differences are also identifiable [4].

Hypermobility of joint was assessed by C-W scoring method: (i) Positioning the thumb to the ventral portion of forearm; (ii) Hyperextension of fingers parallel with dorsal portion of forearms; (iii) Hyperextension of elbow > 10° (iv) Hyperextension of knee > 10° and (v) Flexion of spine with legs extended, and palms resting on the floor.

Beighton score - hyper mobility measuring tool in children. In a study, 35% of school going children scored 5/9 in which 12.3% had joint pain. 9.1% complained of pain after exercise or sports. Goniometry is used to measure (ROM) and joints mobility can also be found in children 6-12 years [5]. Measuring the ROM and getting the normative values helps to interrupt with the abnormality in children on scientific basis of therapeutic intervention [6].

Usually the ROM, changes with male and female which may be connected with joint mobility. Mobility may also be related with age, as age increases joint mobility decreases. Majority

of people score from 0,1 or 2 score in mobility (Beighton and Horan,1969 therefore concluded that the scale is valid for joint mobility measurement [7].

Pain after exercise adds to be noted point for hyper mobility in children under 13 years [8].In the orthopaedic setting greater awareness and diagnoses of hypermobility will lead to better assessment of patients with conditions such as multidirectional instability of shoulder, recurrent dislocation of patella and ankle. The inherent laxity of the connective tissue in such cases may significantly affect the outcome of surgical repair [9].

Hypermobility is not commonly seen among young people but it have to be noted down by the clinicians and need to be treated, as it may be one of the differential diagnosis for rheumatic disease [10]. Synovial joints are most affected by the hyper mobility and is recognized as feature of heritable disorder of the connective tissue and can be identified mostly by the beighton scale [11].

Aim of the Study

The aim of the study is to find out the value of hyper mobility in down's syndrome.

Need of the Study

Only few studies are there to find out the hyper mobility in normal children Hyper mobility of down syndrome where not found before.

Early diagnosis helps Down Syndrome children to prevent hyper mobility

Early diagnosis helps in early intervention program.

Methodology

Study Design : Non experimental study

Study Type : Observational

Sampling Method : Convenient sampling

Sample Size : 30 Subjects

Study Setting : Maithree, National Institute for Empowerment of Persons with Multiple Disabilities, Rehabilitation education center at SRM Hospital (kattankulathur).

Inclusion Criteria

Both males and females were included

Age limit - 6 to 12 years .

Willing to participate in this study.

Exclusion Criteria

Recent fractures congenital anomalies.

Recent surgeries.

Subjects who are not willing will be excluded for the study.

Any deformitie

Procedure

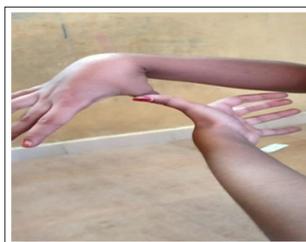
In this study 30 sample were included with above criterias and an inform concern was obtained from the respective parents ,

care taker after the detailed explanation. Hyper mobility is tested using Beighton score, 9 point score requires, Performance of five techniques, four self technique (bilaterally) and one active unilateral performance. So, the selected children were evaluated for hyper mobility using beighton score. Among which the child scoring maximum of 5 out of 9 points were included in the study and the range of motion was checked accordingly.

The Nine Point Beighton Score of Hypermobility

Explanation	Two Sided Testing	Score
Passive DF of the 5th MCP joint to go degrees	yes	2
Passive Hyper extn of the elbow 10°	yes	2
Passive Hyper extn of the knee 10 °	yes	2
Passive apposition of the thumb to the Flexor side of the FA, while shoulder is flexed go degrees ,elbow is extended,and hand is pronated	yes	2
Forward flexion of the spine ,with the knees straight,so that the hand palms rest easily on the floor	no	1

Self positioning of the thumb parallel to forearm, elbow should be extended and hand on pronated position. score +ve if the whole thumb touches ventral aspect of the forearm (Two sided testing).



Score: Positive



Score: Negative

Forward bend with legs straight and touches the floor, If +ve indicates the hand palm rest easily on the floor.



Score: Positive



Score: Negative

Scoring: one point for each side for item (1-4 maximum two per item if left and right are positive) and one point in total for 5 Items. The maximum hypermobility score is nine points (if all items are positive)

Table 1: Passive DF of the MCP. Positive if greater than 90 degree (Bilateral Testing)

Test position	Motion tested	Positioning Goniometer	Anatomical landmarks	Method
Short Chair Sitting Arm - 80 degree Abd, EF- 90 degree, FA resting on table forearm pronated.	Passive dorsiflexion digit 5	MCP 5,	Dorsal metacarpals 5 in the length of digit 5	Lateral method

Table 2: Passive Hyper Extn of Elbow- +ve score > 10 (Bilateral Testing)

Test position	Motion tested	Positioning Goniometer	Anatomical landmarks	Method
Sit on chair with shoulder 90 degree Fb-Flex, FB supinated	Passive Hyper extn of elbow	Lateral epicondyle humerus	Humerus pointed at tub Major humeri Radius pointed at proc styloideus	Lateral method

Table 3: Passive Hyper Extn of Legs. Score positive if > 10 (Bilateral Testing)

Test position	Motion tested	Goniometric point	Anatomical landmarks	Method
Lying backward with legs in horizontal position	Passive Hyperextension of knee	Lateral Femur Epicondyle	Femur pointing to Trochanter and Fibula pointed at lateral malleoles	Lateral method

Outcome Measures

Beighton scale
Goniometry



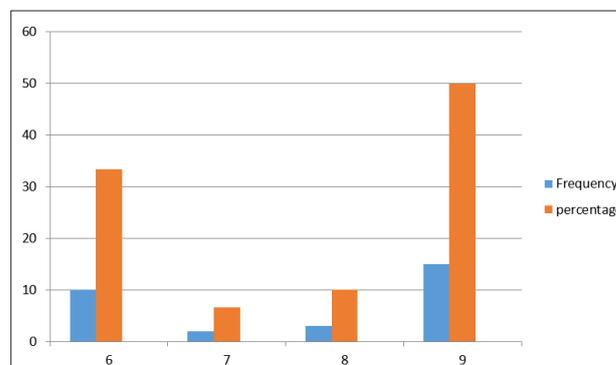
Data Analysis

Table 1: Hypermobility Score with Frequency and Percentage

Hypermobility score	Frequency	Percentage score
6	10	33.3
7	2	6.7
8	3	10
9	15	50

Table-1 : Minimum score of hypermobility was 6 and maximum score of 9 from the benign score of hypermobility. The frequency of hypermobility with 6 score was 10 and percentage score of 33.3, and the maximum score of 9 hypermobility with frequency of 15 and percentage score 50.

**Graph 1
Hypermobility Score with Frequency and Percentage**



Results

Table -1& Graph -1 show the Minimum score of hyper mobility was 6 and maximum score of 9 from the benign score of hyper mobility. The frequency of hyper mobility with 6 score was 10 and percentage score of 33.3, and the maximum score of 9 hyper mobility with frequency of 15 and percentage score 50.

Discussion

Down's syndrome is a third copy chromosome 21, associated with impairment in growth development, IQ, Clinical significance.

Joint hypermobility is one of the hidden feature in many children. Especially in Down's syndrome though hypermobility is commonly found but not evaluated much. Hypermobility syndrome (HMS) is a connective tissue disorder that affects the overall muscular system of the body with stages of severity, leading to secondary problems in future [12].

In general, girls have hyper mobility compared to boys, however in this study also proved that girls are hypermobile than boys with age group of 6-11 years. 33.3% of children showed score of 6 in beighton hypermobility score and 50% of children scored 9 points.

KirkJH has concluded that specific joint hypermobility in patients is considered to have ligament laxity and noted pain during various activities such as long distance walking later on turned to hypersensitivity nerves and weaker immune system.

Recognition of hypermobility children itself is a challenge to all physical therapists, as usually it resolves in later stages of childhood. So, the physical activity need not be restricted unless it's a over flexible based sports such as gymnastics, dance etc.

Goniometry also adds point to the study to find out the degree of motion usually measuring goniometry for hypermobile joints the value exceeds than normal children, which needs to be validated. So in this study hypermobility score as well as goniometric measurements of the joint has been evaluated.

Analysis hyper mobility score value and goniometry measurements showed there is joint hyper mobility among Down's syndrome children and needs care. Children with hypermobility can be given functional training accordingly while early identification and diagnosis may help the physical therapist to plan for the intervention program. In case of hesitation and negligence of hyper mobility among Down's syndrome can lead to further problems that may affect the activities of daily living [13-26].

Conclusion

The study concludes that children with Downs syndrome is found to have joint hypermobility and needs early identification and diagnosis to improve their activities of daily living.

Limitations

Smaller sample size.
No long term follow up has been studied.

Recommendations

Long term effects are recommended. Larger sample size is recommended. Samples can be taken in different centers. Exercises protocol may be recommended in further studies.

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