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Lever Arm dysfunction of Hips in Cerebral Palsy

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Abstract

In cerebral palsy, hip dysfunction is the second most common problem after equines deformity. Abnormal skeletal development of the proximal femur due to abnormal forces in cerebral palsy leads to lever arm dysfunction by reducing the length of the lever arm contributing to one of very important causes of gait abnormality. High hip anteversion, high neck shaft angle, subluxation, and dislocation are the most common lever arm dysfunctions at the hip joint. This lever arm dysfunction increases progressively with age and correspondence to GMFCS level. If not identified at a very early stage then it may progress to completely deform the femoral head & hip joint. Early identification and preventive surgery help in preventing progression in GMFCS I to III but can progress despite of preventive surgery in non-walker children GMFCS IV & V. Recurrence of the problem is also very common in non-walker children. So we need detailed information about the lever arm problem in the hip and how to deal with this problem in cerebral palsy children. In this article, we have discussed etiology, and how to identify and management protocols for different lever arm dysfunctions of the hip in cerebral palsy.

Keywords: Cerebral palsy, Lever arm dysfunction of hip, Hip subluxation, Anteversion

Introduction

Primary abnormalities in cerebral palsy are muscle tone abnormality, poor selective motor control, abnormal movement, muscle weakness and poor balance [1, 2]. This, in turn, leads to bony torsion and muscle shortening. The normal movement of a joint complex is the product of the muscle force and the lever arm (distance of the muscle from the joint insertion). The inability to provide adequate torque leads to lever arm dysfunction and associated gait problems [2]. The lever arm system of the hip joint is a class 1 Lever. In this lever arm, the effort and the load are on opposite sides of the fulcrum. The body weight is the load; the hip abductor force, primarily supplied by gluteus medius, is the effort; and the hip joint acts as a fulcrum. Hip rotation deformity, high neck shaft angle, subluxation, and dislocation are the most common lever arm dysfunctions at the hip joints [2]. For correction of gait abnormality, we need to correct lever arm dysfunction so that muscles can work with their full effort. After equines, hip displacement has been regarded as the most common deformity in children with CP [1]. In this article, we will discuss important issues that affect the lever arm function of the hip and the methods to manage them.

Lever arm dysfunction: Hip in cerebral palsy child can have three types of lever arm dysfunction:

- 1. Short lever arm:** In the coronal plane, the valgus position of the femoral neck shortens the perpendicular distance between the greater trochanter (point of force application for abductors) to the centre of the femoral head (fulcrum) resulting in the generation of the lesser magnitude of internal abduction moment [1, 2]. Often hip abductor cannot meet the demand of increase hip abduction forces so sway develops.
- 2. Malrotated lever arm:** In the axial plane, increased femoral anteversion also

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compromises the lever arm of the hip abductors due to the posterior position of the greater trochanter, placing the hip abductor at a mechanical disadvantage [2]. Therefore, internal hip rotation is a compensatory phenomenon that brings greater trochanters to the frontal plane and increases the lever arm of abductors [3].

3. Unstable fulcrum: Subluxation and dislocation of the hip joint give rise to instability, resulting in an unstable fulcrum. The generated force will not be able to transfer through unstable fulcrum [1,2].

4. Positional lever arm problem: Fixed flexion deformity of the hip leads to a positional lever arm dysfunction by preventing full hip extension during gait [1].

Aetio-pathogenesis

Femoral neck anteversion (FNA): Increased femoral anteversion is one of the most commonly seen transverse plane deformities in children with cerebral palsy. It is the angle between an imaginary transverse line crossing to the knee joint from the medial to the lateral side and the other line from the centre of the femoral head to the base of the neck passing through the centre of the femoral neck [2]. In the new-born, the average FNA angle is 31 degrees and reaches 15 degrees by 16 years of age. But in cerebral palsy, this remodelling does not happen [4]. Increased femoral anteversion in CP may represent a failure of remodelling of the foetal anteversion. Femoral neck anteversion is also known as either femoral ante torsion, medial femoral torsion, internal femoral torsion, or persistent foetal alignment [4]. Reduction in femoral neck angle occurs during growth in normal developing children. It is also believed that upright posture and full hip extension contribute to the remodelling of foetal anteversion in able-bodied young children. The majority of corrections occur up to the age of 5 years. As the hip comes into full extension during standing and walking, the anteverted femoral head and neck come into contact with the anterior Iliofemoral (Bigelow's) ligament. The torsion stress from being in contact with the Bigelow's ligament produces a rotational force on the femur. As children with CP are usually late to walk and often do not achieve a fully upright posture with full extension at the hip, it is probable that they fail to remodel the proximal femur [3]. Apart from this, spastic medial hamstrings and adductors overpower antagonists, thus creating torsion stress. This torsional stress can lead to deformities in growing bones. Sitting posture in an extreme position of internal rotation, or "W sitting," also creates forces that prevent correction of anteversion. Internal rotation gait is not always because of increased hip anteversion but can also occur because of the dynamic component, including spasticity of an internal hip rotator, increased plantar flexor spasticity, and as compensation for weak hip abductor [5].

High femoral Head shaft angle/femoral neck-shaft angle: It is a

common belief that the femur undergoes twists due to rotational forces applied at a right angle to the growth phase of the epiphysis. Heuter Volkmann Law of Epiphyseal Pressure states that an increase in pressure throughout the epiphysis reduces growth, whilst a drop in pressure increases growth. Elastic connective tissue, abnormal muscle forces, and abnormal posture can produce changes in femoral neck angle [4]. An increase in anteversion, neck shaft angle, and migration percentage is directly correlated to GMFCS level [4].

Hip subluxation / Dislocation: A hip "at risk" for instability has a migration percentage of >30%, while a migration percentage of >50% is associated with an increased risk of hip dislocation. Hip subluxation in CP is an acquired condition due to muscular imbalance, persistent femoral anteversion, coxa valga, delayed weight bearing, and abnormal muscle forces [4, 6]. It is suggested that the weaker hip extensors and abductors are overpowered by stronger hip flexors and adductors [6]. As a result, the femoral head centre translates distally to a lesser trochanter, changing the direction and magnitude of the force and thus causing a serious lever arm dysfunction [7]. As a result of this imbalance and proximal femoral and acetabular pathology, most hips dislocate in a posterior and superior direction. Scoliosis also contributes by causing pelvic obliquity, and usually, the hip with the adducted posture on the higher side subluxated [7]. Acetabular dysplasia also develops as a consequence of the pressure of the femoral head to the labrum and migration of the centre of rotation of the femoral head [7]. Root et al. [6] in over 2000 CP children found that quadriplegics have the highest percentage of Subluxation (38%) along with dislocation (15%). On the other hand, hemiplegic, diplegic, and monoplegic have lower rates of 9.5 % and 1% subluxation and dislocation. Brendon Soo [8] mentioned in his population-based study that the incidence of hip dislocation in cerebral palsy was 35%. He found a nearly straight-line relationship between the rate of hip displacement and GMFCS level. He found hip displacement at 15 % in GMFCS 2, 41% in GMFCS 3, 69% in GMFCS 4, and 90% in GMFCS 5. The key finding of his study was that all hypertonic motor types (spastic, dystonic, and mixed) were associated with a similar incidence of hip displacement. The dislocation was associated with pain, discomfort, as well as difficulty positioning, and subsequent development of kyphosis [6]. Unilateral hip dislocation is often linked with pelvic obliquity and scoliosis. Most common acetabular deficiencies are found in the posterolateral direction, but deficiency can also occur anteriorly or globally [10].

Femoral head Deformity: Persistent subluxation and dislocation in cerebral palsy lead to deformity of both the femoral head and the acetabulum. A saucer-shaped deformity

develops in the acetabulum, usually in a cranio-lateral direction, and the femoral head gradually migrates proximally and may also then become deformed [10]. Medial flattening of the femoral head can occur due to excessive pressure on the medial aspect after the proximal migration of the femoral head. The hip capsule and abductors may exert pressure on the lateral side of the head, causing it to flatten. The combination of medial and lateral flattening can produce a triangular or dunce cap appearance to the head, as seen on an anteroposterior radiograph. Along with this deformity, grooves can develop on the superior aspect of the femoral head due to the pressure of reflected head of the rectus femoris [10]. The end result of the femoral displacement is often loss of articular cartilage and early osteoarthritis. High preoperative migration percentage (MP) and long duration of dislocation have been considered to be an important factor for femoral head asphericity [4, 5, 11].

Physical examination: The problem in cerebral palsy is usually at a multi-level, so systemic and thorough evaluation of the child is mandatory before planning for any intervention. The topographical, neuromuscular, and GMFCS classification has to be established, and a functional assessment must be made [1]. Radiology of the hip joint in correct position is very important to assess the stability and position of the femoral head [7]. Correct measurement of hip anteversion is possible by CT scan [7]. All the findings should be correlated with gait analysis [1, 3, 5]. The final check should be done during the per-operative examination [1].

Hip surveillance: Critical early indicators of progressive hip displacement are identified and monitored by Hip surveillance. Early detection is a vital component of the approach to the prevention of hip dislocation and its consequences [10]. It includes clinical assessment and measurement of migration percentage (MP) on the Pelvis AP view radiograph at specific regular intervals, depending upon age and GMFCS level [10].

Treatment: When it comes to managing musculoskeletal deformity, proper treatment of lever arm dysfunction can result in considerable gait improvements. Thus, an appropriate knowledge of lever arm dysfunction in CP gait is of essential relevance when contemplating surgical intervention.

Preventive Surgery —

Soft tissue balancing - Adductor, hamstring, psoas

Guided Growth Modulation: Percutaneous epiphysiodesis by transphyseal screw (PETS) of proximal medial femoral physis

Reconstructive surgery:

Derotational osteotomy

Varus derotational proximal femoral osteotomy

Combined osteotomy, including Femoral and pelvic

osteotomy

Open versus close reduction of hip joint

Salvage surgery:

Proximal Femur resection

Interposition arthroplasty

Valgus femoral osteotomy

Total hip replacement

Preventive surgery: Preventive surgery refers to different surgical strategies to prevent or treat early hip displacement in younger children, whose hip is still in the joint but shows signs of being “at risk” [9]. When the angle of abduction is less than 45, and the Migration percentage is more than 40, then there is a high risk of hip dislocation in future. MP of more than 33% is taken as the threshold for intervention or increased observation and MP of more than 40% taken as consideration of surgery for hips. In children with MP 33–40%, treatment should be based on other clinical signs and the progression of MP over time [12]. The utmost goal of surgical intervention is to correct lever arm dysfunction so that functional deterioration can be prevented and realistic, specific functional goals can be achieved [3]. Soft tissue balancing and growth modulation of proximal medial femoral physis are two preventive surgical measures.

Soft tissue surgery: Soft tissue hip surgery is indicated in patients with cerebral palsy, who have less than 30-degree abduction and MP of 25% to 40%. Adductor release, including adductor longus and gracilis, should be done on both sides in order to equalize the sum of moment of forces at hip level [9, 13, 14]. Partial release of adductor brevis is required in some cases where abduction is less achieved. The neurectomy of the second branch of the obturator nerve should only be done in patients without the possibility of walking and for hygiene purposes only [13]. Psoas muscle may not need release in GMFCS 1-3 children because, it is believed that they can cause weakening of hip flexor power. But if required, then psoas lengthening at pelvic brim is preferable [15]. In children with GMFCS 4 & 5, Psoas tenotomy at a lesser trochanter can be done without any negative impact [10, 15]. Benjamin J Shore [14], in his research, found that in long-term follow-up, the surgical success rate of soft tissue surgery for children at GMFCS level II was 94% compared with 14% for children at GMFCS level V. But it can be one of the temporary measures in GMFCS 4 & 5 to delay the need for reconstruction surgery till the child is medically fit [9, 14]. High success rates are found in ambulatory diplegic spastic children with MP <40% [15]. Every child, who has undergone soft tissue surgery requires close monitoring till the hip is stable for at least six years post-surgery [9].



Figure 1: Screw epiphysiodesis medial physis proximal femur

Guided Growth modulation: Guided growth modulation of the proximal femur can be one of the best options in the early stage of hip subluxation (MP 30-50%) with a high head shaft angle $>155^\circ$ [16]. It is always combined with soft tissue release to get good long-term outcomes [16, 17]. Percutaneous epiphysiodesis using transphyseal screws (PETS) is a minimally invasive outpatient procedure with less comorbidity & gives faster recovery than varus osteotomy [16]. In the literature, the minimum age for this procedure is four years [16]. Earlier growth modulation will give better correction but can be done in a child with two years remaining growth. Best place to put a screw in the infero-medial aspect of the neck in the AP view & central in the lateral view [17]. Growing off of the screw is one of the complications that need regular follow-up and commitment to revision or retightening of the screw when necessary [17]. After removal of the implant following hip reconstruction, guided growth of the proximal femur can be used to minimise the chance of recurrence [17]. Growth modulation of normal-looking hips can also be utilised during reconstructive surgery on the abnormal side. (Fig. 1)

Hip Reconstruction: Kenneth Huh [18] suggests a stepwise surgical approach for hip subluxation and dislocation in children with cerebral palsy. His approach is to do varus derotational osteotomy and release of soft tissue first. Then, he

recommends doing an assessment using fluoroscopy and an arthrogram. If reduction and femoral head coverage are not adequate, then proceed with open reduction and/or pelvic osteotomy. Combined femoral and pelvic osteotomies, along with soft tissue releases, have the most predictable radiological outcome with a lower rate of recurrence compared to VDRO alone in patients with high MP, especially GMFCS level IV or V [6, 18, 19]. Hip asphericity improves with the combined procedure. Preoperative MP, reduction quality, and acetabular coverage influence femoral head remodeling [11]. Even hips with femoral head irregularity can also be candidates for surgical reconstruction, since the risk of subsequent symptomatic arthritis appears to be small [10, 19]. Although some hips can be treated with only a varus osteotomy, adding the peri-ilial pelvic osteotomy will have better results without recognized morbidity if acetabular dysplasia is present. An isolated VDRO was more likely to promote acetabular development postoperatively in patients of GMFCS levels I, II, and III compared with patients of GMFCS levels IV and V. GMFCS level and age of surgery is the predictive factor for postoperative acetabular remodeling after an isolated VDRO [20]. (Fig. 2)

Derotational osteotomy femur: Femoral rotational osteotomy may be performed anywhere proximally or distally with a similar outcome [21]. Proximal osteotomies can be done in intertrochanteric or sub-trochanteric areas. Distal osteotomies are most commonly done at the supracondylar region. Diaphyseal osteotomy is usually preferred in adults and can be fixed by intramedullary nails. Rigid fixation leads to earlier weight bearing and quick recovery. Recovery after distal osteotomy is faster with fewer complications [21]. Usually, the site of osteotomy is based on the associated problem at the hip or knee. Before planning for derotational osteotomy of the femur, contributing factors, including pelvic rotation and hip internal rotator, should be taken into consideration [5]. Derotational osteotomy is only considered when high anteversion is associated with in-toeing or hip dislocation [21]. Recurrence is higher if anteversion is corrected at a younger age <10 years [5].

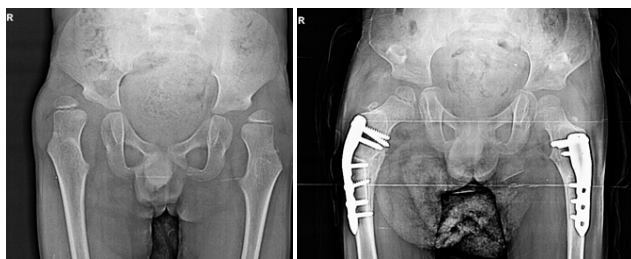


Figure 2a: B/L Hip Dislocation

Figure 2b: B/L Femoral varus & acetabular osteotomy



Figure 3a: B/L hip Subluxation

Figure 3b: B/L Varus osteotomy proximal femur

Varus proximal femoral osteotomy: Hip subluxation of more than 50% MP will progress to dislocation if not intervened, so they require proximal femur osteotomy to reorient the femoral head in the acetabulum. The most common consensus is a neck-shaft angle of 90° to 100° for non-ambulatory children (GMFCS levels IV and V) and 120° for ambulatory children (GMFCS levels I, II, and III) [22]. John M Mazur recommends achieving a 90° angle in less than four years, 100-110 in 4 to 12 years of age, and 110-120 in children of more than 12 years 23. Chances of a second surgery are high if surgery on the hip has been done at less than 4 years of age [24]. So soft tissue balancing surgery and growth modulation are more favourable for quadriplegic children, especially GMFCS 4 & 5 less than 4 years. It will give time for major surgery and better medical conditions for the child. The addition of pelvic osteotomy can help in getting similar results even with less varus correction (NSA angle 130) in the proximal femur and also reduces the amount of adductor release [10, 25]. (Fig. 3)

Acetabular osteotomy: Acetabular dysplasia is very common in patients with persistent subluxation of more than 50% MP and dislocation for a longer duration in quadriplegic. The chances of remodelling are less after the age of 8 years, so it is better to reorient acetabulum after this age [20, 23]. Dega osteotomy is the most preferable pelvic orientation osteotomy in cerebral palsy irrespective of age [7, 11, 22]. Dega osteotomy can correct all posterior and anterolateral defects, which is very common in cerebral palsy. Scott J Mubarak [26] has added bi-cortical cuts at the posterior and anterior corners (the sciatic notch and the anterior inferior iliac spine) of the osteotomy to allow bending of the acetabulum. The remaining cut is unicortical, extending down to the triradiate cartilage. Pemberton osteotomy is only indicated in anterior dislocation with anterior acetabular dysplasia. Chiari osteotomy is indicated only in patients with fused triradiate cartilage and less-than-congruous spastic dislocations [7].

Open versus close reduction: Most cases do not require the opening of the hip joint. In cerebral palsy, soft tissue is rarely an obstacle to reduction, even in severe subluxation as well as

dislocation [24]. If after varus shortening osteotomy of the proximal femur, close reduction is not possible, then open reduction and clearance of acetabulum are required [24].

Unilateral versus Bilateral: There is always controversy regarding unilateral versus bilateral hip reconstruction. Bilateral surgery is preferable in GMFCS IV & V even if the contralateral hip looks radiographically normal [19, 27]. Unilateral reconstruction surgery, along with soft tissue surgery on the opposite side, can be done in children beyond the age of 8 years. But need of close monitoring of the opposite hip till the skeletal maturity is mandatory [27]. The concept of Growth modulation can also be utilised on the opposite side in mild affection. Bilateral surgery can be done simultaneously in one setting [28] or two settings, with intervals of a few weeks [19] depending upon the general condition of the child, the surgeon's experience and the facilities available to them.

Salvage surgery: Salvage procedures are only advisable in cases where the hip has symptoms of arthritis and is also unreconstructable in order to achieve reasonable abduction and pain relief. This includes head and neck resection (Girdle stone procedure), castle interposition arthroplasty, prosthesis interposition arthroplasty, valgus femoral osteotomy, a combination of valgus osteotomy and proximal femoral excision, hip fusion, and total hip replacement [6, 7, 9, 10, 29].

Conclusion

Lever arm dysfunction of the hip is one of the very common problems in cerebral palsy. The prolonged existence of lever arm dysfunction in the hip can induce arthritis of the hip, causing discomfort and interfering with everyday tasks. With early identification, we can slow down the progress of dysfunction. Once this lever arm dysfunction reaches an advanced stage, then we need reconstruction. We should be very watchful in non-walker children till the maturity. To plan the correct intervention, we need to understand lever arm dysfunction in detail and also the criteria for treatment protocol.

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