Surgical Management of Hip Problems in Myelomeningocele: A Review Article

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Abstract

Background: Children with myelomeningocele (MMC) develop a wide variety of hip deformities such as muscle imbalance, contracture, subluxation, and dislocation. Various methods and indications have been introduced for treatment of muscle imbalances and other hip problems in patients with MMC but there is no study or meta-analysis to compare the results and complications. This review aims to find the most acceptable approach to hip problems in patients with MMC.

Methods: MEDLINE was searched up to April 2015. All study designs that reported on the outcomes of hip problems in MMC were included. From 270 screened citations, 55 were strictly focused on hip problem in MMC were selected and reviewed.

Results: Complex osseous and soft tissue reconstructive procedures to correct hip dysplasia and muscle balancing around the hip are rarely indicated for MMC patients without good quadriceps power.

Conclusion: Over the years a consensus on the best algorithm for treatment of hip dislocation in myelomeningocele has been missing, however, muscular balancing with/out osseous procedure seems a reasonable approach especially in unilateral mid-lumbar MMC.

Keywords: Hip dislocation, Hip dysplasia, Myelomeningocele, Meningomyelocele, Spina bifida, Teratology

Introduction

Myelomeningocele (MMC) is a saclike structure containing cerebrospinal fluid and neural tissue, caused by a failure of the neural tube to close during the fourth week of gestation (1). It is a common malformation, occurring in approximately 1 in 1,000 live births. The cause is unknown, but, both genetic and environmental factors have significant roles. MMC may be located anywhere along the neuraxis, but, 75% occur in the lumbosacral region (2). In the 1960s, effective techniques were developed for shunting hydrocephalus and early closure of neural tube defects. As a result, orthopaedic surgeons were presented with the challenge of managing a population of children who had MMC (3). Initially, the musculoskeletal problems in these children were treated with the modalities that had been learned from the treatment of poliomyelitis. However, it soon became apparent that the management of children who have MMC was not so simple because of additional factors such as decreased sensation of the lower extremities or encephalopathy that impair coordination and results in the loss of strength of the lower and upper extremities and muscle imbalance that affects skeletal development over the entire period of growth (1-3). Progressive neurological deterioration may occur in MMC because of tethered cord syndrome, syringomyelia or hydrocephalus (4, 5). Hydrocephalus in association with a type II Arnold-Chiari defect is common and develops in 80% of children with MMC but only 30% of patients need a neurosurgical shunt (2, 6). Ambulation in patients with MMC is affected not only by the neurological level of motor performance, but also by other factors such as age, obesity, cognitive status, motivation, spasticity, upper limb functional status, orthopedic deformities and intelligence (7, 8). At minimum follow-up of 20 years 42% of MMC had normal IQ but only 8% achieved college degrees (6).

Materials and Methods

We reviewed articles concerning different treatments of hip problems in MMC. The search engine was MedLine (PubMed) and the keywords used were:
Until April 2015, 270 articles were retrieved for review. Articles were included if they provided an abstract and were written in English. Of those, 55 were selected and reviewed when they were strictly focused on the topic of this article. In addition, references of these articles were examined to ensure no compelling literature had been overlooked. The types of studies reported have a low level of evidence (most of them are level III-IV studies).

Results

Classification

The neurological level of lesion is an essential factor influencing ambulation in children with MMC and hip problem (9). The most frequently used classification systems of the neurological level of lesions in the literature are Sharrard (1964), Hoffer (1973), Lindseth (1976), Ferrari (1985), Mcdonald (1991), and Broughton (1993)(7).

Hoffer and colleagues’ classification was based on studies of 56 patients with MMC aged between 5 and 42 years. The resulting classification system has four categories: thoracic, upper lumbar, lower lumbar, and sacral. Also they considered four levels of functional ambulation consisting of community ambulators, household ambulators, non-functional ambulators and non-ambulators (7, 8). In 1999, Bartoneck analyzed these six commonly used classification system in patients with MMC and suggested that it is not possible to compare neurological levels in different classification system to each other or definitely correlate the results to the functional ambulation levels of Hoffer classification (7, 8).

A modification of the classification system described by Asher and Olson based on the lowest level of antigravity (at least grade-3) muscle strength on the patient’s best side to define the neurological level is simple to use, and has been helpful in predicting gross motor function and potential problems (3, 10).

Thoracic level: Patients who have only functional thoracic roots may be able to walk during the first decade of life, but, they become dependent on a wheelchair as they attain adult body mass. They do not have active movement of the lower extremity muscles and have some abduction, external rotation, and flexion contracture around their hip (11-14).

Upper lumbar level (L1, L2): They have active flexion of the hip but seldom retain the capacity for functional walking after reaching the adulthood. A contracture of the unopposed hip flexors typically develops in these patients. A mild contracture of the unopposed hip adductors may also occur, however, restriction in hip abduction is usually mild and is not clinically important(3, 12).

Mid-lumbar level (L3, L4): Patients who have mid-lumbar MMC typically have normal strength in the hip flexors and adductors but no function in hip extensors or abductors. Therefore, a flexion contracture of the hip and some limitation of abduction frequently develop in these patients. More importantly, this pattern of muscle imbalance predisposes to progressive subluxation of the hip (8).

Lumbosacral level: The probability of subluxation or a severe flexion contracture of the hip is low in patients who have lumbosacral MMC. This is particularly true if the MMC is at the sacral level, but adequate stability and an adequate range of motion of the hip are usually maintained even in patients with fifth lumbar MMC who have only grade 2 or 3 strength of the hip abductors and absent or trace strength of the hip extensors (15, 16). Apparently, this degree of activity of the hip abductors combined with activity of the hamstrings can be an effective counterbalance to hip flexors and adductors of normal strength. These patients should have periodic radiographs during early childhood to monitor the development of the hip joint. It was previously thought that all patients who had sacral MMC were able to walk independently; However, in a study, among 36 patients who were evaluated as adults, 6 had become dependent on a wheelchair as a result of neurological deterioration, ulceration of the feet, and other problems (15).

Natural history: Neurological level is a critical indicator in determining the ambulation capacity, functional capability and hip deformity. 56% of adults with MMC are unemployed and 43% use wheelchair (6). All patients in L2 level are wheelchair dependent and 60% of patients below L2 level use wheelchair sometimes. Maximal level of ambulation will achieve at age four to six years old and if the child cannot stand at six years old then walking will be impossible (16).

Samuelsson et al. studied factors determining ambulation in 163 patients with MMC by a multivariate statistical method and concluded that severe scoliosis was closely, age was moderately, and hip flexion contracture was slightly related to the inability to walk, while pelvic obliquity, hip dislocation, or knee flexion contracture were not (17). Also several other studies have demonstrated that the ability to walk is not affected by dislocation of the hip in patients who have thoracic or upper lumbar MMC (11-14, 18-21). Therefore, complex osseous and soft tissue reconstructive procedures to correct hip dysplasia and muscle balancing around the hip are rarely indicated for these patients since relocation of the hip with a good radiograph does not mean functional advantage and may cause complications, such as pathological fracture or devastating stiffness of the hip joint (6, 22). Although

<table>
<thead>
<tr>
<th>Level</th>
<th>Function</th>
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<tr>
<td>Thoracic</td>
<td>No grade-3 strength in muscles of lower limbs</td>
</tr>
<tr>
<td>L1-L2</td>
<td>Hip flexion or Adduction</td>
</tr>
<tr>
<td>L3</td>
<td>Knee extension</td>
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<td>L4</td>
<td>Knee flexion</td>
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<tr>
<td>L5</td>
<td>Ankle dorsiflexion</td>
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<td>S1</td>
<td>Ankle plantar flexion</td>
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Patients with MMC can also be functionally classified in four groups: (3)
Posterolateral transfer of the ILP

In 1952 Mustard transferred external oblique muscle and femoral the greatest average value of flexion contracture was in the hip in older children (9-11 years old) revealed that not correct (41). In a multicenter study of 1061 patients does not occur; however, nowadays we know that this is no muscle activity around the hip then hip dislocation in limbs without any innervation below T12 and no transfer of iliopsoas to the greater trochanter (39, 40). Sharrard (1964). He introduced the posterolateral muscle imbalance around the hip in MMC began by hip extensors. The story of the surgical treatment of iliac window to the greater trochanter, but Mustard's introduced lateral transfer of the ILP through the anterior to allow bracing (3, 38).

Up to 80% patient with MMC have a problem or weakness in upper extremity and Charney found that, with bracing and gait training, 52% of 87 patients who had MMC at upper level were able to walk about the community at age five but deficient balance reaction and weakness of the upper extremities coupled with the extensive bracing that is needed, prevents some of these children from achieving a functional walking ability (16, 36). Children who have upper level MMC rarely retain the ability to walk after reaching the adulthood and hip dislocation in MMC is not painful compared to CP, so, some authors have advocated an intensive program of bracing and gait training during early childhood. The potential benefits of walking by patients with MMC at upper level include strengthening of the upper extremities, protection against obesity, and prevention of contractures of the lower extremities. However, the most important benefit is the psychological boost and tremendous sense of accomplishment that these children express when they achieve the ability to be upright and to move around a room like other children of their age (37, 38). Furthermore, in a study, patients with this level of MMC who were managed with bracing and early walking had fewer fractures and pressure sores and were more independent in transfers, even when they eventually switched to a wheelchair compared with patients who had always used a wheelchair. However, these children were hospitalized more often, for operative procedures were hospitalized more often, for operative procedures to allow bracing (3, 38).

Muscle balancing procedures: In 1952 Mustard introduced lateral transfer of the ILP through the anterior iliac window to the greater trochanter, but Mustard’s procedure was abandoned because it did not reinforced hip extensors. The story of the surgical treatment of muscle imbalance around the hip in MMC began by Sharrard (1964). He introduced the posterolateral transfer of iliopsoas to the greater trochanter (39, 40). Also. He found no hip dislocations or flexion deformities in limbs without any innervation below T12 and no active muscles around the hip. He said that if there is no muscle activity around the hip then hip dislocation does not occur; however, nowadays we know that this is not correct (41). In a multicenter study of 1061 patients with MMC, measurement of the flexion contractures of the hip in older children (9-11 years old) revealed that the greatest average value of flexion contracture was in the patients who had thoracic or upper lumbar MMC. Dislocation tended to occur by the age of three to four years in the patients who had mid-lumbar MMC, but those who had thoracic or upper lumbar MMC continued to have dislocation of the hip even after the age of ten years (18).

Time of transfer: Posterolateral transfer of the ILP should be limited to a selected group of patients. Sharrard reported some deformities of patients, presented at the age of one year after birth in 183 children with MMC, so, in 1983 he recommended the transfer should be done before development of osseous deformity (between one and two years of age), combined with an adductor release, and should be limited to patients who have fourth lumbar MMC (29). Also later two studies demonstrated the Sharrard procedure or external oblique muscle transfer before age one is unsuccessful. In the study by Stillwell and Menelaus, five out of nine patients with ILP transfer before one year old, were not able to walk during the follow-up, at least ten years postoperatively but they provided useful data concerning the effectiveness of the Sharrard procedure in other ages (30). Also, Tosi et al. transferred external oblique muscle and femoral osteotomy after initial treatment with a Pavlik Harness and reported redislocation of two of four hips. The reason for the redislocation was unclear, but the early operation, or early wrong grading of quadriceps may explain them (31). Because early grading of quadriceps power in first 3 years of life give a reliable assessment of future ambulatory ability, but it should be noted that early grading of quadriceps power compared to grading at later ages, only 56 out of 109 assessments remained the same (42).

Absence of flexor power after ILP transfer and role of flexion contracture around the hip: Stillwell and Menelaus have reported 47 patients with ILP transfer and adductor release to obtain 60 degrees of abduction more than 10 years ago among which, 32 (68%) were community walkers, 3 were household walkers and 12 were non-walkers. Compared to other published reports, these patients did not lost their walking ability that could be jeopardized by the loss of hip ILP flexor power. Furthermore, all except three of the community walkers were able to climb and descend stairs after ILP transfer (30).

Many patients with intact flexor and absent extensor function around the hip developed hip flexion contractures. Contracture in household ambulatory patients ranged from 0 to 45 (average 23) degrees. As hip flexion contracture did not appear to impair the ambulatory ability when the reciprocating gait orthosis was used, the surgical release of functioning hip flexor musculature if the use of this device is being considered, is not indicated and up to 30 degree of the hip flexion contracture is acceptable (16, 41, 43).

Shurtleff et al. analyzed 5,147 serial measurements of the range of hip extension in 966 patients with spina bifida and concluded that the contractures were generally present in the first few months of life (physiologic flexion posture); this then diminished during the first 27 months in all but those with thoracic lesions. They demonstrated that the contractures reappear or worsen between the ages of 3 and 6 years and are not merely due
to muscle imbalance, sitting posture, or these factors in combination. Hence, surgical management is seldom appropriate until after that age (44). Also Frowley showed that successful isolated anterior hip release is not related to the neurological level or the operation age; also, recurrence of contracture correlated with the walking ability of the child at the latest follow-up (43).

**Muscle balancing procedure should be done in located hip or subluxed hip?** Based on the fact that muscular imbalance of the hip is responsible for progressive hip dislocation and secondary osseous deformity, Weis et al. (1988) reported ILP transfer on 54 hips; 23 of them (42.6%) had located hips at the moment of the operation (32). Molloy (1986) combined the ILP transfer with femoral varus derotation osteotomy on 26 hips; five hips (19.2%) were located and did not have dysplastic changes (33). Bunch and Hakala (1984) performed an ILP transfer on 32 hips, but three of them (9.4%) showed no abnormal changes (28). Lorente (2005) performed the ILP transfer on 24 hips, 41.4% of hips were located in his series (45). However, it is now understood that progressive dysplasia does not develop in all patients who have MMC at the mid-lumbar level, and experienced observers, such as Broughton and Menelaus, after a review of 1061 patients, concluded that muscular imbalance is not a significant factor in the production of flexion deformity or dislocation of the hip and they do not recommend prophylactic surgery. They recommend a selective approach for operative intervention only for patients in whom subluxation has developed when have at least grade-4 strength of the quadriceps and 90 degree hip and they do not recommend prophylactic surgery.

**ILP or external oblique transfer:** It is now clear that posterolateral transfer of the ILP does not provide active extension or abduction against gravity, and it is doubtful that this out-of-phase transfer provides any noticeable extension or abduction during the gait cycle (3). The transferred ILP or external oblique muscle is unlikely to provide significant active abduction or extension power, however ILP transfer may have a beneficial function as a tenodesis (31, 46). In our limited experience some abduction power is achieved after ILP transfer only in supine position. A recent study using three dimensional gait analysis revealed no improvement in abnormal pelvic obliquity in patients with fourth lumbar MMC who had been managed with ILP transfer (47). Transfer of the external oblique muscle has been advocated as an alternative to transfer of the iliopsoas for patients who have MMC at the mid-lumbar level as well as a dysplastic hip (31, 43, 48). This procedure does not weaken the iliopsoas; therefore, the power of the hip flexors and the ability to climb stairs should be maintained. Some authors have stated that transfer of the external oblique muscle improves hip mechanics during mid-stance, however, gait-analysis studies have demonstrated that

the transferred external oblique muscle mainly functions during the swing phase of gait therefore it is doubtful that this transferred muscle simulates the activity of the hip abductors or extensors during walking (35). In a comparative study external oblique muscle transfer did not provide a clinically important improvement in functional recovery in patients with L3 to L5 level when added to periarticular release of contractures and bony procedures (49). In spite of these articles it is difficult to compare the results of ILP and external oblique muscle transfer because a comparative study is not yet done between two transfers in MMC and there is not any general agreement on the choice of the muscle transfer (30, 31, 34).

**Triple transfer:** Phillips and Lindseth (1992) described the results of triple transfer of the external oblique muscle to the greater trochanter, the hip adductors to the ischium and the tensor fasciae latae to a more lateral position on 89 hips (34). Although those authors reported functional walking by all patients, the duration of follow-up was not enough.

**Transfer in bilateral dislocation:** According to Menelaus (1980) and Caroll (1987), due to leg length discrepancy, pelvic obliquity develop, resulting pressure sores, poor sitting balance, and a negative influence on the status of spinal column, bilateral dislocations should be treated only when full power in quadriceps muscle exists and unilateral dislocation should be treated regardless of the level of paralysis (50), but Robert (1994), Crandall (1989), and Sherk (1991) have reported that the incidence of these problems with the exception of a leg length discrepancy is not significantly influenced by unilateral dislocation of hip and they do not recommend the procedure in inappropriate set (19).

**The role of osseous procedure in combination with muscular balancing procedure:** Muscle transfers for the treatment of dysplasia of the hip associated with MMC are insufficient to correct severe osseous deformities. Misalignment of the proximal part of the femur or the acetabulum is particularly common after the age of three years and this problem should be corrected either before or at the same time as the muscle transfers (3). A femoral varus derotation osteotomy corrects abnormal valgus angulation and anteversion. The type of pelvic osteotomy that best serves these patients is less clear. These patients often have global acetabular deficiency (51). For that reason, a Pemberton or modified Dega procedure may be better than other pelvic osteotomies that are commonly used for the management of young children who have CDH (3). Chiari osteotomy did not achieve long-term hip stability and good results in many patients (52, 53). In a study of 34 children (66 hips) with third, fourth, or fifth lumbar MMC who had a femoral osteotomy combined with transfer of the external oblique and adductor muscles, Tosi et al. reported a maintenance of stability of 57 (73%) of 51 hips in the 26 children who remained neurologically stable; however, only eight of 15 hips in children who had progressive loss of neurological function remained stable (31). The poorest results were for the hips that had dislocated previously. Only two out of 10 hips in this group had
successful results. The average duration of follow-up in that study was relatively long (10.9 years), but the wide range of follow-up (0.7 to 20.0 years) limits conclusions concerning functional status when those children reached adult body sizes. At the most recent evaluation, 21 (81 per cent) of 26 children who did not lose neurological function during follow-up were able to walk about the community. So even after ossseous procedure in MMC, progressive loss of neurological function is an important factor for the stability of hip and ambulation. The combination of one stage pelvic and femoral osteotomy and transiliac psos transfer can be effective in selected patients with MMC.

Discussion

Treatment in the first year of life: As mentioned above, muscle transfer has poor outcomes before the age of one year; so, treatment of subluxation or dislocation of the hip in midlumbar MMC is difficult during the first year after birth. Typically, the problem is noted in the first few months of life. Use of a Pavlik Harness or some other brace designed for CDH is seldom successful over the long term for these patients (3). Furthermore, these braces exacerbate a flexion contracture in an infant when the hip extensors are nonfunctional (3). Based on Green (1998) and Breed (1982), when dysplasia develops in the first year of the life in patients who have midlumbar MMC, it develops by the age of three or four months and many of these patients probably have dislocation of the hip at birth, but the treatment of other medical problems prevents its documentation. If intervention is delayed, the rapid growth during infancy coupled with an underlying muscle imbalance may result in severe dysplasia that cannot be stabilized with soft-tissue procedures. Because pavlik can induces flexion contracture of hip and is not successful to treat muscle imbalance, they recommend an ILP recession and adductor myotomy (3, 54). They recommended early surgical treatment to prevent secondary adaptive changes in the hip (54).

Treatment of progressive subluxation of the hip in adolescent patient: Progressive subluxation of the hip in adolescent patient with midlumbar MMC presents a dilemma. Muscle imbalance coupled with complicated hip dysplasia leads to a substantial rate of recurrent subluxation. There is no answer for this group of patients. Certainly, the evaluation should include assessment for a possible tethered cord syndrome and consideration of a CT scan with reconstruction to define the extent and location of the acetabular deformity (51, 55). Base on Instructional Course Lectures AAOS (1998), the treatment must be individualized (3). Green have most often performed a Chiar pelvic osteotomy and femoral varus derotation osteotomy in this group of patients. He have also recommended observation or no treatment for adolescent patients who have a history of operations, no functional abductor muscles, and a markedly dysplasic acetabulum. In this situation, the chance of success is low and reconstructive operations may cause the hip to become stiff and painful (3). However Mannor (1996) and Zenios (2012) showed Chiari osteotomy did not achieve long-term hip stability and good results in MMC (52, 53).

References

8. Hoffer MM, Feiwell E, Perry R, Perry J, Bonnett C. Functional ambulation in patients with...


