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Teratologic Hip Dislocations : Controversies and Consensus

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Abstract

Teratologic hip dislocations are rarer and more severe than their typical developmental dysplasia counterparts. Due to a diverse array of etiologies and associated comorbidities, management of teratologic dislocations is challenging. Prognosis following the treatment of teratologic hips is therefore guarded.

The current literature on the management of teratologic hips has enabled us to arrive on some common consensus. Unilateral hips should be aggressively treated, while bilateral hips need careful evaluation. Closed reduction is universally discouraged. Open reduction, with or without bony procedures, is the standard treatment. These cases also require prolonged bracing, extra care during perioperative period, and long-term follow-up. Multiple surgical procedures and complications, both immediate and delayed, are commonplace in teratologic hips. Apart from medical and surgical interventions, there are many non-modifiable factors which determine the prognosis of these cases. This truth has to be accepted by the health-care team and family members to develop a realistic expectation toward functional outcomes following treatment.

Keywords: Hip dysplasia; Arthrogryposis multiplex congenita; Varus derotational osteotomy; Pavlik harness; Hip spica.

Introduction

Teratology is the study of abnormalities of physiological development [1]. Teratologic hip dislocations are those that occur in conjunction with severe and pervasive underlying diseases of neuromuscular etiology. These hips show marked abnormality at the time of birth [2]. It is evident that the pathology and the consequent anomalous development of the hip start early in fetal life. This stands in sharp contrast to typical developmental dysplasia of the hip (DDH) which occurs as an isolated entity [1]. The latter is characterized by hips which are in various grades of dysplasia but is quite supple and without joint contractures or stiffness.

In this review article, we will shed more light into the comparatively lesser known type of hip dislocation. We will also attempt to build a consensus by reviewing the current literature on this entity.

Incidence and Etiology

In contrast to the typical DDH, which is fairly common, teratologic dislocation of the hip (TDH) is a rare disorder. Its incidence is reported to be 0.03-0.04/1000 births [3].

Teratologic hip dislocation can occur in many conditions. Arthrogryposis multiplex congenita is by far the most common entity. As described by Bamshad and Hall, it is the common phenotypic feature of more than 300 specific disorders [4, 5]. These include neuromuscular disorders, skeletal dysplasias, multiple congenital contracture syndromes, and aneuploidy. Incidence of hip involvement in arthrogryposis ranges from 55 to 90% [6]. Other conditions where teratologic hip dislocation can occur are Larsen syndrome, diastrophic dwarfism, Goldenhar syndrome, neural tube defects, Hallermann-Streiff syndrome, absent vertebral segment, chromosomal abnormalities, and mucopolysaccharidosis [7].

Question 1: Include neural causes?

There is some controversy regarding inclusion of neural causes as teratologic etiologies for hip dislocation as paralysis of the lower limbs may present a different set of issues. Similarly, cases with excessive ligamentous laxity were excluded from the list of teratologic causes [8]. However, because of the fact that arthrogryposis multiplex congenita itself includes many disorders which are predominantly neural in origin, it would be safe to argue in favor of inclusion of these entities into the spectrum of disorders causing TDH provided their effect starts early in utero.

Consensus 1: Neural causes which are operative since early fetal life should be included.

The exact mechanism of teratologic hip dislocation is unknown [2]. The intimate and persistent contact between the femoral head and the acetabulum is important for the development of normal anatomy of the hip joint in utero. It is postulated that the lack of contact between the two components of the hip joint leads to gross distortion of the anatomy of teratologic hips. Katz suggested that embryonic insult or primary germplasm defect with consequent muscle imbalance may be responsible for this dysplasia [1].

Question 2: What are the clinical and radiographic differences between TDH and DDH?

The pathological findings are much more pronounced than a non-syndromic hip: The acetabulum is small; the roof is oblique or flattened. Fibrofatty tissue fills up much of the cavity. The ligamentum teres may be thickened. The femoral head is distorted, with the medial side being flattened. The dislocation is usually anterior even though the version of the neck is variable ranging from high degree of anteversion to retroversion. The surrounding soft tissues are abnormal and the muscles are often poorly developed [6, 7].

These pathological changes translate into corresponding clinical effects on physical examination. The hip is stiff and irreducible. The hip is severely and rigidly displaced such that it may not be possible to perform an Ortolani test [2]. The greater trochanter may be felt proximally, signifying a high dislocation. A general examination reveals typical findings of a more pervasive generalized disorder in the body. Akazawa enumerates the cardinal features of an arthrogrypotic child: (1) Joint contracture at birth in at least two different areas of the body; (2) evidence of a non-progressive neurological disorder; (3) diffuse muscle wasting with fusiform joint degeneration; (4) generalized reduction in muscle bulk, shortening of muscles with decreased power, and skin dimpling over a contracted joint; (5) the presence of webbing; and (6) the absence of normal skin creases [9]. The child may display a "wooden doll" appearance. In diastrophic dysplasia, the typical findings are short-limbed dwarfism, "hitchhiker thumbs," multiple joint contractures, and a normal skull [10]. In neural tube defects, there will be varying grades of muscle power in the lower limbs, sensory deficits, and incontinence. Examination of the back will reveal swelling, dimple, or a tuft of hair in the midline.

3. Open reduction by anterior approach: To circumvent the problems of difficulty in reduction and preventing stiffness and redislocation, Akazawa et al. performed extensive anterolateral approach by removing all the obstacles to a successful reduction including attachments of rectus femoris, iliopsoas, gluteus medius and minimus, and short external rotators [9]. He performed a complete circumferential capsulotomy followed by release of joint contractures. According to him, a high dislocated femoral head is compressed against the iliac wall and the posterior capsule is adherent to the ilium. Akazawa also stated that incomplete release of the tight structures and adherent capsule may in fact lead to more stiffness and subluxation than a more radical release. However, a radical release risks damaging femoral head perfusion. In spite of meticulous attempts to preserve the blood to the hip through the lateral epiphyseal artery, 70% of cases experienced avascular necrosis of the femoral head. However, the range of movement was significantly improved and all children could walk independently.

Radiographs in a TDH show a dislocated hip (Fig. 1a), more often a high dislocation. Acetabular dysplasia is variable.

Consensus 2: Patients with TDH present with more severe and advanced clinical features than their typical DDH counterparts.

Management of the hip in teratologic pathology is difficult [2, 3, 6, 7]. Multiple joints are often involved and many of them need surgical interventions. However, the child is less able to tolerate major surgical procedures due to multiple comorbidities. There are many non-modifiable factors which determine the walking ability of the child and in spite of the best procedures and meticulous rehabilitation, it is often difficult to assure significant improvement. A large proportion of cases require multiple procedures to maintain an enlocated hip. In that attempt, many hips lose their blood supply, range of motion or become painful. The following are the treatment issues merit further discussion.

Question 3: How should we treat Unilateral TDH?

Unilateral hips have to be treated aggressively regardless of ambulatory ability [11, 12, 13]. This is because Coleman et al. found that fixed pelvic obliquity compromises sitting balance even in non-ambulators and makes bracing and ambulation more difficult. According to Drummond and Mackenzie, such pelvic obliquity resulting from unilateral hip dislocation contracture may produce scoliosis [14].

Consensus 3: Every attempt should be made to reduce a unilateral hip dislocation.

Question 4: What is the role of Surgical intervention in bilateral TDH?

Treatment is more controversial in bilateral dislocations because of the lower probability of obtaining two hips that remain reduced and supple [15]. Different schools of thought surround bilateral teratologic hip dislocations. The first school advocates conservative approach to the bilateral dislocations as they are usually neither unstable nor painful [7, 12, 13, 16]. Bilateral rigid dislocated hips have stiffness as a persistent issue even after surgery [7, 12, 13]. Greater range of motion preoperatively favours a good outcome after surgery. Coleman suggested that anterior dislocations may often be left untreated because they cause little shortening and do not produce much adduction contracture [11].

The other view is to reduce all dislocations to restore normal hip biomechanics and decrease the risk of future pain or stiffness [9, 17].

St Clair and Zimbler tried to form a consensus on this by dividing children with bilateral dislocations and arthrogryposis into two main groups: (a) For patients with severe stiffness of the hips, severe muscle weakness, and severe involvement of the upper limbs, a low ambulatory potential may be expected, and reduction of dislocation is not recommended; (b) for children with greater mobility of the hips and less involvement of upper limbs, a good ambulatory potential may be expected and reduction should be attempted [18].

Consensus 4: Surgical intervention in bilateral cases is to be attempted at least in those who have good ambulatory potential.

Question 5: Which surgical method is successful in TDH?

1. Closed reduction: Unlike an isolated dysplasia where conservative treatment is the preferred management in early infancy, closed reduction of a teratologic dislocation is not advocated. Closed reduction has been shown to have low rates of success [1, 2, 3, 6, 7, 8, 9]. It may lead to avascular necrosis, spurious reduction, deformation of the femoral head, stiffness, and repeated surgical procedures [7].

2. Open reduction by medial approach: Staheli, in his series of 18 patients, treated with medial open reduction found

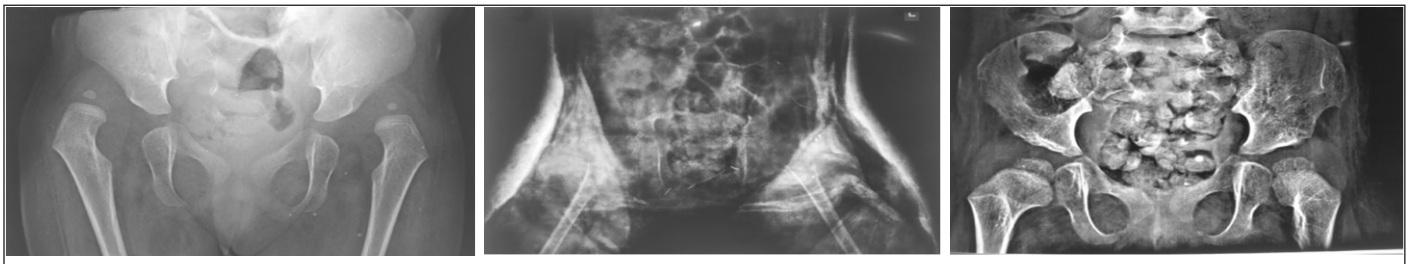

Figure 1: Illustrative case of Teratologic Hip Dislocation. Female child born with bilateral hip dislocation, bilateral knee dislocation and bilateral telipes equinovarus. a) Shows high dislocation of both hips. b) She was treated with bilateral open reduction through anterior approach and immobilized in hip spica. c) Hips at the age of three years. She is ambulatory at home with bilateral ankle foot orthoses.

that range of motion was better than with other methods [17]. Interestingly, his patients were younger (mean age at surgery was 9.7 months). It is generally agreed that utility of such procedures may be limited to low-grade dislocations and in children younger than 18 months [6].

3. Open reduction by anterior approach: To circumvent the problems of difficulty in reduction and preventing stiffness and redislocation, Akazawa et al. performed extensive anterolateral approach by removing all the obstacles to a successful reduction including attachments of rectus femoris, iliopsoas, gluteus medius and minimus, and short external rotators [9]. He performed a complete circumferential capsulotomy followed by release of joint contractures. According to him, a high dislocated femoral head is compressed against the iliac wall and the posterior capsule is adherent to the ilium. Akazawa also stated that incomplete release of the tight structures and adherent capsule may in fact lead to more stiffness and subluxation than a more radical release. However, a radical release risks damaging femoral head perfusion. In spite of meticulous attempts to preserve the blood to the hip through the lateral epiphyseal artery, 70% of cases experienced avascular necrosis of the femoral head. However, the range of movement was significantly improved and all children could walk independently.

4. Open reduction and bony procedure: Huurman and Jacobsen reported better functional results using sub-trochanteric extension osteotomy [19]. It is generally agreed that femoral shortening is helpful in the reduction of the high femoral head in AMC or in congenital dislocation of the hip. Many of the subsequent papers are replete with relative success of such procedures in addressing dual issues of stable reduction without compromising blood supply.

Consensus 5

Closed reduction should not be attempted in TDH. Anterior open reduction is a good option, with supplemental bony procedures where necessary. Medial open reduction is a viable alternative in younger patients. Extensive releases should be performed bearing in mind the vascular anatomy of the femoral head.

Question 6: Single event or staged procedures?

Le Bel et al. proposed a 3-stage approach in complex cases [2].

1. Open adductor and psoas tenotomy followed by 2 weeks skin traction;
2. Open reduction of the hip and spica cast for 6 weeks.
3. Varus derotational osteotomy of the proximal femur was performed at the end of treatment.

With this approach, the authors achieved independent ambulation in 9 out of 13 patients, while 8 patients required additional surgical procedures. The AVN rate was 20%. These results are comparable to those obtained through single event surgery.

Consensus 6: Staged surgery may not offer additional benefit.

Question 7: What is the best age for correction of TDH?

Williams suggested that feet be corrected at 4 weeks of age, knees at 8 weeks, and hips at age 6–8 months [6, 20]. However, surgery should be performed only when the general condition allows the child to safely tolerate major intervention. Additional procedures, most commonly for acetabular coverage are deferred till the age of 4-5 years. By this time, the potential for ambulation as well the course of the underlying disease is more evident.

Consensus 7: Hips should follow correction of the foot and the knee at around walking age and in accordance with the surgical fitness.

Question 8: What precautions should be taken postoperatively in TDH?

Because soft tissues surrounding the hip are abnormal, there is a high risk of recurrence if bracing is weaned too early. The hip spica usually remains in place for about 12 weeks. According to Huurman and Jacobsen, it may be necessary to use a brace at night for several years to attain maximum remodeling. Since the head and acetabulum may already be deformed at the time of surgery, remodeling to congruency may take many months to occur [6, 9].

Consensus 8: A long period of bracing is important.

Question 9: What are the complications encountered during the treatment of TDH?

Complications of surgical interventions are frequent and have been mentioned in every case series. Some complications are directly related to the surgery, AVN being the most significant. AVN may occur due to injury to the vessels during overzealous release around the hip or

due to attempts at forcible reduction. AVN can cause proximal femoral deformity, stiffness, and limb length discrepancy. Redislocation or subluxation is another concern, and results from deficient coverage, poor positioning, muscle imbalance, or compromised reduction. Many of the hips may be rendered stiffer after the procedure.

Children with TDH have poor general health and multiple comorbidities. Infections have frequently been described in such children [2]. Osteoporotic fractures, allergic reaction to sutures, and post-operative cardiopulmonary arrest have been described in the literature. They also present with unique perioperative challenges [21, 22]. Venous access may be difficult due to poorly developed vasculature. Endotracheal intubation is problematic because of their small stiff jaws. The tendency to develop intraoperative hyperthermia is also high. Distribution of anesthetic drugs may be significantly altered due to decreased muscle mass. They also have higher incidence of atelectasis, aspiration, and stridor during the post-operative period. Children with skeletal dysplasia may have unrecognized CV junction anomalies and positioning during intubation requires prior evaluation of such a possibility.

Consensus 9: Complications are common and should be considered during management.

Question 10: What is the prognosis for children with TDH?

Gibson and Urs stated that the prognosis was relatively poor for children with affected hips: 25% are confined to a wheelchair, 25% are dependent on braces for walking, and only 50% can walk independently [13]. However, those children who are cognitively normal usually develop compensatory mechanisms to get around their impairments. Moreover, a small improvement in range of motion, stability, and comfort improves their quality of life significantly [6].

Consensus 10: Although prognosis is variable following treatment of TDH, some improvement in quality of life can be expected and treatment is justified

Discussion

From this literature review, the points of agreement may be summarized as follows:

The clinical, radiological, and pathological findings of teratologic hip dislocation are more pronounced than those of DDH. The hips have high dislocation, joint contractures, dysplastic muscles, and soft tissues.

Most authors discourage closed treatment. In unilateral cases, aggressive surgical reduction is advocated. In bilateral cases, the chances of obtaining symmetric hip reduction and containment have to be weighed against the risks of recurrence, complications, stiffness, and repeated surgeries. An anterior or anterolateral approach can address all the aspects of the problem including release of impeding structures (Fig. 1b), wide release of the capsule, femoral shortening, and provision of acetabular coverage. Medial open reduction is an option in the younger age group with low dislocations. In case of multiple joint involvement, the foot, knee, and hip are successively managed in that order. The perioperative management of the arthrogrypotic patient can be difficult and is best handled by an experienced anesthesia and medical team.

A considerable period of bracing is required for the soft tissues to settle. While a contained painless and supple hip allowing independent community ambulation is the ideal goal, a painless hip with preserved range of movement should be the first priority of every health-care provider concerned with the care of the patient. In the end, it is the realistic expectation of the family members and joint decision with all the caregivers which has the best possibility of producing outcomes satisfactory to all.

Conclusion

Teratologic hip dislocations represent some of the most severe types of hip dysplasia. They inherently have more marked clinical and radiological findings and have a tendency to resist treatment, especially those employing conservative and minimally invasive methods. These children deserve treatment consisting of diligent decision making followed by whole-hearted surgical procedures if indicated, steadfast follow up and long term support to achieve meaningful outcome.

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