

ORIGINAL ARTICLE

CONTROVERSIES IN CONGENITAL CLUBFOOT : LITERATURE REVIEW

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Despite common occurrence, congenital talipes equinovarus (clubfoot) is still a subject of controversy. It poses a significant problem with its unpredictable outcome, especially when the presentation for treatment is late. The true etiology remains unknown although many theories have been put forward. A standard management scheme is difficult as there is no uniformity in the pathoanatomy, classification and radiographic evaluation. These differ according to the age of the child and the severity of the condition. The paper discusses these controversies with an emphasis on the proposed etiologies and types of treatment performed.

Key words : congenital talipes equinovarus, controversies, management.

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Introduction

Congenital talipes equinovarus (CTEV) or clubfoot is a common condition. In Malaysia, owing to the ignorance of parents, clubfoot remains a significant problem and yields an unpredictable outcome due to late presentation for treatment. Clubfoot deformity was documented as early as ancient Egypt. Smith and Waren in 1924 found that Pharaoh Siptah of the XIX Dynasty was afflicted with clubfoot (1). Talipes Equinovarus was first introduced into the medical literature by Hippocrates in 400 B.C.V. (1, 2). He recognized that some clubfeet were congenital, while some were acquired in early infancy. The term talipes equinovarus is derived from Latin: talus (ankle) and pes (foot); equinus: "horse like" (the heel in plantar flexion) and varus: inverted and adducted. Hippocrates (1) also suggested that the treatment should start as soon as possible after birth with repeated manipulations and fixations by strong bandages which should be maintained for a long time to achieve over correction. His teaching principles of treatment are as valid today as they were 2300 years ago.

Incidence

The incidence of clubfoot varies widely with race and sex. The overall incidence of clubfoot was 1 to 2 per thousand live births (3,4). The incidence in the United States is approximately 2.29 per 1000 live births (5); 1.6 per thousand live births in Caucasians (6); 0.57 per thousand in Orientals; 6.5 to 7.5 per thousand in Maoris (7); 0.35 per thousand in Chinese; 6.81 per thousand in Polynesians (8) and as high as 49 per thousand of live births in fullblooded Hawaiians (9). Boo and Ong (10) reported the incidence of clubfoot in Malaysia 1.3 per 1000 live births. Males outnumber females by 2:1 with 50% of cases being bilateral (6). In those with unilateral deformity, there was a right sided predominance (11). A higher incidence of clubfoot was also noted in patients with a positive family history (3, 6).

The possibility of clubfoot occurrence in a sibling was 1 in 35 and if present in an identical twin, the risk was 1 in 3 (12). Although this was probably due to polygenetic influences, it was suggested that it might also be due to an autosomal dominance of poor penetrance (13).

Etiology

The true etiology of clubfoot remains unknown. Many theories have been put forward:

1. Mechanical factors in utero

This is the oldest theory and was first proposed by Hippocrates (1, 2, 11). He believed that the foot was held in a position of equinovarus by external uterine compression. However, Parker in 1824 and Browne in 1939 believed that diminution of amniotic fluid, as in oligohydramnios, prevents fetal movement and renders the fetus vulnerable to extrinsic pressure (2).

2. Neuromuscular defect.

Some investigators still maintain the opinion that equinovarus foot is always the result of neuromuscular defect (14-17). On the other hand, others have shown no abnormalities in their histological studies (19-20) and electromyographic studies of the muscles in clubfoot (21, 22).

3. Primary germ plasma defect.

Irani and Sherman (23) had dissected 11 equinovarus feet and 14 normal feet (18). In clubfoot, they found that the neck of talus was always short, with its anterior portion rotated medially and plantarly. They suggested that the deformity probably resulted from a primary germ plasma defect.

4. Arrested fetal development

a) Intrauterine environment

In 1863, Heuter and Von Volkman first proposed that the arrest of fetal development early in embryonic life was a cause of congenital clubfoot (2). This theory was maintained by Bohm in 1929 (21, 22). However, the opponents of this theory were Mau (1) and Bessel-Hagen (2).

b) Environmental influences

The harmful influence of teratogenic agents on fetal environment and development are well exemplified by the effect of rubella and thalidomide in pregnancy. Many authors believe that there are various environmental factors responsible for the

appearance of a clubfoot, as there are various substances capable of producing a temporary growth arrest (21, 24, 25).

5. Hereditary

Clubfoot tends to be familial in a significant number of cases (6, 9, 19, 26). It is inherited as having a polygenic multifactorial trait (6, 10, 13, 24, 26). Wynne-Davis stated that polygenic inheritance is more susceptible to the influence of environmental factors (6).

Pathoanatomy

Numerous anatomical studies of clubfoot have confirmed the gross changes in the shape and position of the talus, navicular, calcaneum and cuboid (18, 27-31). The tendons, tendon sheaths, ligaments and fascia of the foot have undergone adaptive changes and became fibrotic or contracted (19, 28, 31-35). The talocalcaneocuboid joints are subluxated (2, 5, 23, 29, 36, 37). Nevertheless, until today, the question still remains as to whether the initial anatomical changes first occurred in the tarsal bones with subsequent soft tissue adaptation, or vice versa.

Classification

The purpose of a classification system is to help in subsequent management and prognosis. Various classifications of clubfoot exist in the literature (1, 3, 38-40). However, without a uniform standard, these classifications pose a major problem. Furthermore, some are too complex for practical use.

Dimeglio in 1991 divided clubfeet into 4 categories based on joint motion and ability to reduce the deformities (39).

1. Soft foot – may also be called postural foot and corrected by standard casting or physiotherapy treatment.

2. Soft > Stiff foot – 33% of cases. It is usually a long foot which is more than 50% reducible and responds initially to casting. However, if total correction has not been achieved after 7 or 8 months, surgery must be performed.

3. Stiff > Soft foot – 61% of cases. It is less than 50% reducible and after casting or physiotherapy, it is released surgically according to specific requirements,

4. Stiff foot – it is teratologic and poorly reducible. It is in severe equinus deformity, often bilateral and requires an extensive surgical correction.

Clinical Features

Congenital clubfoot must be differentiated from postural and structural or secondary type of clubfoot. The postural clubfoot has the clinical appearance of congenital clubfoot, but it can become fully correctable to normal anatomic position at birth, or shortly thereafter following a period of manipulative strapping. The patient should be thoroughly examined to exclude features of paralytic clubfoot including multiple congenital malformations.

Radiological Assessment

At present, there are no satisfactory methods for an early objective assessment. In 1896, Barwell introduced the use of plain radiographs to assess the exact status of clubfoot (1). However, at birth, clinical examination is more informative than radiological assessment, as only the ossification centres of the talus, calcaneum and metatarsals are present. These two tarsal bones appear as small rounded ossicles. Thus, the plain radiograph film does not help to evaluate the shape and orientation of the tarsal anlage. The tarsal bones become sufficiently ossified after 3 to 4 months. By then, radiological evaluations give a more accurate objective record than does clinical evaluation. Some authors have made radiological assessments by an anteroposterior and lateral projection films before and after surgical correction. (2, 11, 37, 41-43). Up to date, there is no consensus on the value of radiographs in the routine management of congenital clubfoot.

Treatment

The management of clubfoot continues to present a formidable difficulty owing to the current views on its pathoanatomy and treatment. The results of any form of treatment vary according to the severity of deformity and the surgeon's philosophy on this deformity.

The aim of treatment is to obtain an anatomically and functionally normal feet in all patients. (42). However, this is unrealistic as the deformity of the joints and ligaments of the foot and

the ankle are sometimes too severe to be corrected fully. Conservative treatment of clubfoot is well accepted and has been reported to result in good correction ranging from as low as 50 % to as high as 90% (42). Recent trends show that gentle plaster manipulation is more popular than strapping. This serves two purposes :

1. Completely correcting the clubfoot as the definitive treatment. Mild clubfoot may fall into this category.
2. Partially correcting a rigid clubfoot thereby making the surgical approach less extensive (44, 45). Casting tends to prevent further tightening of the contracted structures during the interval prior to surgery (44). The treatment should be started early as the earlier the treatment is started, the easier and better the outcome of results are (46,-48). It will allow preservation of the articular cartilage, optimal growth of the bone particularly talus and maintenance of joint mobility (49).

Manipulation

Manipulation should be gentle but yet strong enough to stretch the soft tissue contractures. Forceful manipulation may result in a spurious correction producing rocker bottom foot. Traditionally as suggested by Hippocrates (2), the components of clubfoot deformity were corrected from distal to proximal (i.e. correction of supination, forefoot adduction and followed by equinus). However, this concept is no longer popular, as equinus, varus and adduction deformities occur simultaneously and not as an isolated component. Thus, attempts are made to correct all elements of the deformities simultaneously.

Following manipulation, an above knee plaster cast is applied with the foot held in maximum correction. While the cast is setting, it is moulded around the heel to lock the calcaneum in the corrected position. The amount of correction must be monitored to avoid compromise to the blood circulation (2, 46, 47). The cast is changed at weekly intervals for the first 6 to 8 weeks, then at fortnightly. Evaluation is done after three months of treatment. If a satisfactory correction is demonstrated, the foot is held in an overcorrected position by a series of plaster cast or an orthotic splint. Dennis Browne splint was popular at one time, but its use had been compounded by a high failure rate of skin irritation, apart from also being cumbersome (2, 50-52).

Below knee casts are difficult to maintain without subsequent slippage in those with significant equinus, extremely small everted heels, chubby legs and short rigid feet.

Unfortunately, some of these deformities recur or become resistant to further conservative treatment. A foot is currently considered resistant when the deformity shows no evidence of further improvement after 3 months of adequate conservative treatment (53). Surgical treatment is inevitable then. However, opinion diverges as to the proper surgical procedure of choice. Argument centers around the nature and the timing of the operation required for the resistant clubfoot. Recent trend towards early soft tissue release between 3-6 months of age is well supported by many authors as sufficient time is allowed for the tarsal bones to achieve maximum remodeling (4, 54-59). There is little evidence that the children who are operated on before 3 months of age have better results. This is owing to the small size foot and the difficulty in differentiating between the tendons and nerves (2, 21).

The general concept in the surgical treatment of clubfoot is to achieve complete and permanent correction with one operation (2, 60). Decision to choose the categories of operative procedures depends on age of patient, degree of rigidity and presence of deformity. The surgical procedures that are currently in use can be divided into three basic groups. These procedures are :

1. Soft tissue procedures
2. Combination of soft tissue and bony procedures
3. Bony procedures.

The procedure that involves soft tissue consist of release or lengthening of tight , deforming soft tissue structures such as ligaments, joint capsules and tendons, as well as performing tendon transfers. The incision used vary widely, but what is performed beneath the skin is far more important to the result than the incision itself. The simplest soft tissue procedure is the posterior release , which involves tendo Achilles lengthening, posterior capsulotomy of ankle and subtalar joints and sectioning of the calcaneofibular and posterior talofibular ligaments, as these ligaments prevent dorsiflexion of the talus (61). The comprehensive soft tissue release include

the posteromedial release of Turco 91 and circumferential release (49, 57, 62). tendon transfer is occasionally performed to provide dynamic balance between the evertors and invertors (63, 64). Magone et al in 1989 reviewed all the three soft tissue procedures done at Columbus Children Hospital and was unable to definitely state which procedure is better (54). Other authors have reported 70%-91% of good to excellent correction on patients underwent posteromedial release before 6 months of age, and 50% relapse rate when this procedure was done after 9 months old (4, 55, 56, 65).

In the child whose tarsal and metatarsal bones have become deformed and resist correction, a combination of soft tissue release and various bony procedures are considered (66-68). In older children between five to eight years of age, a combination of soft tissue release and Lichtblau procedure (resection of distal end of calcaneum) is recommended (69, 10). In those older than nine years of age, the lateral column of the foot is shortened and stabilized by calcaneocuboid resection and fusion (66). A combination of soft tissue release with a medial opening wedge osteotomy of calcaneum and insertion of a bony wedge is also described (71).

In general, bony procedures are rarely if ever, indicated in the infant and young child as these will disturb the normal growth and development of the foot. In a skeletally mature foot (more than ten years old), osteotomy of the os calcis, tarsal reconstruction and triple arthrodesis are required as salvage procedures (3, 72). Metatarsal osteotomy at their bases will correct the varus footfoot, Dwyers osteotomy of the calcaneus corrects hindfoot varus (70, 73) and medial rotation osteotomy of the tibia may be indicated to correct severe lateral rotational malalignment of the tibia and fibula (74). Occasionally, a talectomy is performed (11).

Conclusion

Congenital clubfoot is still a subject of controversy and remains a significant problem owing to the unknown aetiology and disputed pathoanatomy. Moreover, there are no satisfactory methods for early objective assessments and consensus on the value of radiographs in the routine management. The results of any form of treatment vary according to early presentation for treatment, severity of the deformity and surgeon's philosophy on the deformity.

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