Congenital clubfoot

How I do it

Ad F M Diepstraten

Department of Pediatric Orthopedic Surgery, Sophia Children's Hospital, Dr. Molewaterplein 60, 3015 GJ Rotterdam, The Netherlands. Tel +31 10-4636363. Fax -4636844

Epidemiology

Congenital clubfoot (talipes equinovarusadductus) is one of the commonest congenital deformities of the lower limb. The incidence is estimated at between 1 in 700–1000 live births in the USA and Western Europe. A higher incidence is found in the Polynesians and a lower in the Chinese. Boys are affected twice as often as girls. Almost half the cases are bilateral.

Pathogenesis and etiology

About 80% of all clubfeet occur in children who are otherwise physically and mentally normal. Despite much research, the exact pathogenesis and etiology remain obscure. It does not seem to be caused by intrauterine injury or germ defect or by neuromuscular disease. The only feature that is well known is heredity: the risk of having another child with clubfoot is close to 10%.

The other 20% of clubfeet are associated with neuromuscular disease, like spina bifida and arthrogryposis multiplex, skeletal dysplasia or congenital malformation syndromes. Even in these cases the pathogenesis is unclear: it is not a consequence of simple muscular imbalance. I will discuss idiopathic clubfoot only.

Pathological anatomy

Most data about the pathological anatomy of clubfeet derive from studies of fetuses or stillborn infants, which may not be representative of the clubfeet seen in daily practice. One intriguing feature of the clubfoot deformity is the great variety of stiffness, ranging from easily correctable to very stiff feet, which do not respond to any manipulation at all. In the classical description (Irani and Sherman 1972) of the pathological anatomy, the whole foot is in equinus in the talocrural joint, the hindfoot is in varus and the mid- and forefoot are in adduction, supination and cavus. The talus is smaller and the talar head and neck deviate in the plantar and medial directions. The capsules and ligaments at the posterior and medial sides of the foot and ankle are thickened and shortened, as also are the tendons, tendon sheaths and muscles. The dislocation of the talonavicular joint is supposed to be the essential lesion.

McKay (1982), Carroll et al. (1978), Simons and Sarrafian (1983) and recently Epeldegui and Delgado (1995) are credited with a more extensive description of the morbid anatomy. There has been a lot of debate about the position of the talus in the talocrural joint and about the presumed malrotation in the skeleton of the leg. McKay (1982) has shown that the tibia and fibula have a normal rotational position in relation to each other, and the axis of the talar body has no abnormal rotational position relative to the transverse axis of the ankle. The axis of the talar neck and head may deviate in the medial and plantar directions. The main abnormality is in the so-called subtalar complex, consisting of the talocalcaneal, the talonavicular and calcaneocuboid joints. The complete deformity of the foot can be explained as a triplane malposition, the interosseous ligament between the talus and os calcis acting as a central pivot. The navicular is localized next to the medial malleolus, leaving the anterior aspect of the talar head uncovered and pointing laterally instead of medially. The talar head can thus be palpated easily on the lateral side of the foot. Whether the position of the joints of the tarsus should be called a dislocation, a subluxation or an extreme malposition is more a semantic than an anatomical question.

An essential part of the description of the pathological anatomy of the clubfoot is the so-called posterolateral tether (Scott et al. 1984). The posterior part of the os calcis is kept firmly close to the tip of the lateral malleolus by a mass of fibrous tissue, consisting of

the fascia of the deep dorsal compartment of the leg, the sheaths of the peroneal tendons, capsules and ligaments of the talocrural and subtalar joints. It prevents the tuber calcanei from moving downwards and medially during dorsiflexion and eversion of the foot.

Physical examination

Once you have seen a clubfoot, it is not difficult to recognize the next one. The feet show varying degrees of stiffness and passive correctability. In almost all cases there is a single deep transverse skin crease over the distal Achilles tendon instead of the usual 4 or 5. Sometimes there is a transverse skin crease at the medial side of the foot. One finds redundant skin over the anterolateral side of the tarsus, where the talar head can be felt. At an attempt to dorsiflex the foot, the tuber calcanei remains in its high position, and therefore the heelpad is empty.

Special attention should be paid to the spine and the hips to exclude congenital neuromuscular disease and hip dysplasia. Congenital malformation syndromes are often obvious. If in doubt, a pediatrician, clinical genetist or pediatric neurologist should be consulted.

There is no need to confirm the clinical diagnosis of clubfoot by radiography. Other congenital abnormalities in the skeleton of the foot are very rare but, when suspected, may require radiography for diagnosis.

Treatment

The objective of the treatment of clubfoot is obvious: one wants to obtain as normal a foot as possible. This means a plantigrade, straight and painless foot with a good range of motion of ankle and tarsus, with balanced muscle activity and without the need to wear modified shoes.

It is agreed that treatment should start soon after the diagnosis. Some authors state that treatment should start on the first day of life, when correction is easier due to tissue laxity induced by maternal hormones.

Almost general agreement exists about starting treatment by closed methods; in only a few centers are clubfeet treated by primary surgery in the neonate (Ryöppy and Sarinen 1983). Whatever closed method is used, one should realize that the soft tissues are, in fact, the hard ones in clubfeet. The soft cartilaginous parts of the skeleton of the foot withstand less easily the forces of manipulative correction than the shortened capsules, ligaments and tendons. The cartilage is easy to deform and may be put under so high pressure that growth may be impaired. The deformed foot should be handled gently.

Different combinations of manipulation, strapping, casting and physiotherapy have been described and each method has its supporters. To my knowledge, no prospective randomized outcome study has compared different closed methods.

Closed treatment is disappointing in the sense that the majority of the feet cannot be corrected completely and that some kind of surgery is often necessary. The next problem is how long the closed treatment should be continued. In Europe, closed treatment is often used for 2–4 months (Bensahel et al. 1990). At that time the surgeon should have made up his mind about the next step in the treatment plan. Many clubfeet look rather good after closed treatment, show a reasonable range of motion, seem to be straight and the parents are often impressed by the result. The decision to treat a particular foot by surgery is mostly made on clinical and radiographic grounds. Only postural clubfeet are likely to be corrected completely on radiographic examination and further treatment does not seem to be necessary. Whether—and which type of surgery is indicated for the other feet seems to depend on the education and experience of the particular surgeon: there is no agreement in the literature about the kind and extensiveness of the surgery to be used. There is, however, a tendency to correct all the residual deformity after closed treatment in one operation: limited and staged surgery is not common today.

There also seems to be a tendency to operate at an earlier age. Most surgeons prefer to have finished the intensive treatment when the child starts walking (Bensahel et al. 1990). Defenders of restricted surgery fear over- and undercorrection, postoperative scarring and stiffness, injury to neurovascular structures, disturbed wound-healing and infection after extensive surgery (Ponseti 1992).

Depending on the extensiveness of the dissection of the foot and ankle, various operations may be distinguished: a simple posterior or posterolateral release (Hudson and Catterall 1994), the popular posteromedial release (Turco 1979) and the peritalar or circumferential release (McKay 1983, Simons 1985). All these operations are soft-tissue releases; it is agreed that operations on the skeleton of the foot and ankle should be reserved for older children or for recurrences. A real recurrence of the deformity is uncommon; most so-called recurrences are probably undercorrections. As in all surgery, secondary operations have a much greater complication rate than primary surgery.

Secondary bony operations are used when soft tissue releases alone are not expected to correct the deformity because of misshaped bones and articular sur-

faces and insufficient time for remodeling by growth. Calcaneal osteotomy may correct heel varus (Dwyer 1963), calcaneocuboid joint resection and fusion may shorten a too long lateral foot ray (Evans 1961), an opening wedge osteotomy of the medial cuneiform bone may elongate a too short medial ray (Hofmann et al. 1984) and a residual adduction of the forefoot can be corrected by metatarsal osteotomies (Berman and Gartland 1971). After the age of 10 years, a triple fusion is often the last resort to correct a stiff, undercorrected varus deformity of the foot. Severe malrotation of the leg requires a supramalleolar osteotomy. Overcorrection of the hindfoot in valgus is even more difficult to treat, because of the often concomitant mid- and forefoot adduction and supination.

Tendon transfers in the leg and foot are mostly used to correct muscle imbalance in neuromuscular disease. They are not so popular anymore for correcting of slight undercorrection of idiopathic clubfeet. Transfer of the posterior tibial tendon in a young child may result in planovalgus deformity of the hindfoot. Lateral transfer of the anterior tibial tendon is still used by some physicians (Ponseti 1992).

After a soft tissue release, performed in the first year of life, cast immobilization for 6–12 weeks is necessary for the healing of the lengthened tendons and ligaments and for stabilization of the reduced joints. It also allows the changed anatomy to accommodate by growth. Too long immobilization will stiffen the foot, too short will lead to loss of correction. After the cast immobilization, most surgeons use some kind of splint or brace to maintain the foot in the corrected position. The enormous variety of devices, static and dynamic splints, to be worn during sleep or around the clock, biomechanically simple or sophisticated, cheap or expensive, means that almost every orthopedic surgeon has invented his own device.

Evaluation and follow-up

It is difficult to compare different treatments of clubfeet because of variations in the severity of the deformities, the ages of the children, and the methods of assessment. The criteria for clinical and radiographic success do not always correlate with the patients judgement, a problem often encountered in medicine. Only a few large outcome studies describe the function of the treated feet in adult life (Laaveg and Ponseti 1980). Short-term outcome studies on closed treatment followed by soft tissue releases performed in the first year of life have been published by numerous authors. Striking and also disappointing is the fact that there is a great variety of successful cases of conservative treatment only, confirming the lack of consensus in pediatric orthopedics regarding the classification at presentation and after closed treatment, indications for specific surgical treatment and clinical and radiographic criteria for evaluation.

My view

My personal view of the treatment of idiopathic clubfeet is based on 10 years of pediatric orthopedic experience in the largest pediatric university hospital in The Netherlands. From 1975 on, about 700 cases of clubfeet have been treated according to a rather strict protocol, which has been revised during these years in details only.

Primary assessment and closed treatment

Primary assessment of neonatal cases is usually performed on the first day of life when the child is born in our hospital or at a convenient time in the first week of life when the child has been born in another hospital or at home. In my opinion, there is no reason to treat clubfeet as an emergency.

After exclusion of other congenital abnormalities and neuromuscular disease by a general physical examination and sometimes by sonography or radiography, closed treatment is started. The correctability of the deformity is assessed. Unilateral, slender and long clubfeet in girls tend to be less stiff then the bilateral short and broad feet in boys. Manipulation of the foot is done gently and according to the principles described by Kite (1972) and Ponseti and Campos (1972). It is important not to try to reduce the deformity completely in the first manipulation, but to reduce forefoot adduction first together with longitudinal traction in an attempt to stretch the shortened medial structures to facilitate the reduction of the navicular to its normal position in the talonavicular joint. A snugly fitted plaster cast well padded above the knee is applied with the foot still in equinus and the knee bent at 90 degrees to prevent the cast from sliding distally. Two 6-centimeter broad plaster rolls are sufficient. The toes of the foot are left out of the cast to disclose quickly a gradual slip of the cast and the parents are instructed to contact the outpatient clinic, in case the toes disappear into the cast. The manipulation and the application of the cast have to be performed by the treating physician himself or under his direct supervision by a resident, and not left to a plaster-technician or a nurse. The manipulation and casting are repeated every week and later every 2 weeks. The cast is removed by means of a plaster shears instead of a powered saw. The latter will scare the infant

as well as the parents and at least once a year one will make a cut in the skin of the child. A more frequent change of the cast is of no benefit; the correction is not only dependent on mechanical stretching of soft tissue, but also on remodeling by growth. Only after correction of the adduction deformity of the forefoot is a reduction of the subtalar complex attempted. This means that the navicular has to be brought in front of the talar head, the cuboid must be brought in front of and in line with the anterior part of the os calcis and the tip of the tuber calcanei put medially in order to derotate the os calcis from under the talus in the lateral direction. By making this quite complicated multiarticulated correction in the tarsus, the hindfoot is brought into valgus automatically. By pushing the heel into valgus by pressure over the medial and posterior parts of the os calcis, one prevents the os calcis from derotating properly. At every manipulation, an attempt is made to stretch the triceps surae and heelcord but the whole foot is not put in dorsiflexion before the other parts of the deformity are corrected. Some authors (Ponseti 1992) advise a percutaneous tenotomy of the Achilles tendon under local anaesthesia, if the heel still remains in a high position after a few manipulations. The goal is to facilitate dorsiflexion of the whole foot in the talocrural joint. I have no personal experience of this early tenotomy and, after many operative corrections of very stiff feet, I do not believe it to be beneficial.

After 5 or 6 changes in the cast, most clubfeet have revealed their true nature. Already now it is possible to predict the future of that particular foot. Most so-called postural clubfeet are corrected completely and in the very stiff feet there seems to be no correction at all in the tarsus. The talar head is still pointing in the lateral direction on the anterolateral part of the foot and the first ray of the foot cannot be positioned laterally in abduction without exorotating the whole foot and leg. From that moment on, no forceful correction of the foot should be performed, because it will lead to spurious correction and damage to the cartilaginous skeleton of the foot. The closed treatment is continued until the age of 3 or 4 months, depending on the size of the foot and the residual deformity. The cast is removed and the passive range of motion in the tarsus and ankle is then assessed. Active contraction of extensors and peroneals is mostly observed only in the well corrected feet.

At that time, standard radiographs of the foot are made. While taking the radiographs, the foot is held by the treating physician. An anteroposterior, or better said, a superoinferior view at an angle of 30 degrees is taken while the foot is held in plantarflexion and maximum abduction of the forefoot and eversion of the tarsus. A precise lateral view of the ankle and tarsus is taken, while holding the foot in maximum dorsiflexion and eversion. For protection of the hands of the person who holds the foot, the foot is kept in the desired position with the aid of a wooden board. The radiographs of a well corrected foot will show a normal alignment of the talus and os calcis. This means that they should diverge on the anterorposterior view and converge, with a small overlap, on the lateral view. The os calcis must be in dorsiflexion on the lateral view. On the anterorposterior view, the axis of the talus has to point medially in relation to the first ray and the cuboid must be in line with the os calcis. The sum of the angles between the axis of the talus and os calcis on both views is called the talocalcaneal index and it should be more than 40 degrees (Figure 1).

A normal relationship of the bones of the foot and a residual equinus of the whole foot on the lateral view mean that there is no need for further correction in the tarsus and these feet need only a simple posterior or posterolateral release. On the stressed lateral view of the foot, very often an overcorrection of the cavus deformity in the midtarsal joint is seen, a so-called rocker-bottom foot (Figure 2). Some authors dislike this view. To me it means that a further attempt to correct the equinus is useless and that the foot needs surgery instead of a splint or cast.

If, on the anterorposterior view, the talus and os calcis do not diverge and the axis of the talus is pointing to the second to fifth rays or even more laterally, and on the lateral view the both bones are parallel and the whole foot is still in equinus (Figure 3), a more extensive soft tissue release is indicated. Thometz and Simons (1993) have paid attention to the possible medial position of the cuboid in the calcaneocuboid joint, which means that the adduction of the foot is localized in the midtarsal rather than in the tarsometatarsal joints. I strongly advise an accurate study of the radiographs; I believe they depict the residual deformity much better than the clinical examination. For a proper interpretation of the views, it is important that the surgeon himself should hold the foot in the right position while taking radiographs. After the decision to operate, one waits some weeks for the skin to recover and the foot to regain motion.

Operative techniques

As mentioned above, a simple posterior or posterolateral release is reserved for the foot that only shows a residual equinus deformity. The child is positioned prone and a tourniquet is used. After a longitudinal skin incision medially along the Achilles tendon, the

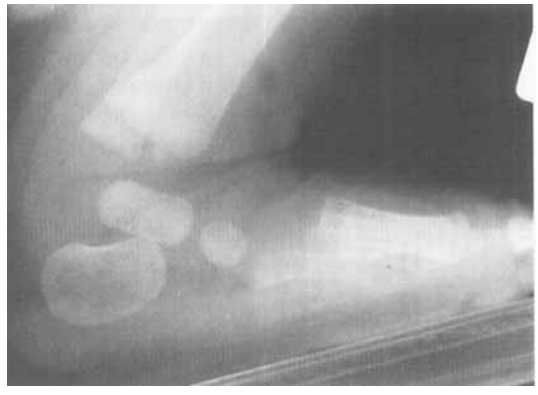

Figure 1. Anteroposterior and lateral radiographs of a well corrected clubfoot at age 4 months. This foot needs no surgery at all.

Figure 2. Anteroposterior and lateral radiographs of a partially corrected clubfoot at age 4 months. This foot needs a simple posterior release only.

Figure 3. Anteroposterior and lateral radiographs of a clubfoot at age 4 months, in which the deformity within the tarsus requires a complete release.

heelcord is lengthened by a Z-plasty. The fascia of the deep compartment is opened by an L-shaped incision, the flap being held medially by a blunt retractor together with the muscle and tendon of the long flexor of the hallux, thus protecting the neurovascular bundle without the need to identify the nerves and vessels. A complete dorsal capsulotomy of the talocrural and subtalar joint is carried out. The capsule is excised in case of severe thickening. I always use a pair of scissors instead of a knife and check carefully whether the instrument is in the joint itself, because it is easy to create a false joint space in the cartilage of the talus by using a knife. An important step in the posterior release is the sectioning of the so-called posterolateral tether. Achilles tendon lengthening and dorsal capsulotomy are not enough to bring the foot into proper dorsiflexion. The fibrous tissue at the posterolateral side of the ankle, including the sheath of the peroneal tendons and the calcaneofibular ligament, prevents the os calcis from moving downwards. In all cases, a careful handling of this tether is mandatory for normal dorsiflexion of the foot. After suturing the heelcord without tension and after hemostasis, the wound is closed and an above-the-knee plaster cast with the foot in maximum correction is applied.

A more extensive soft tissue release is indicated in the feet which, on clinical examination, cannot be put in valgus and abduction and on radiographic examination show an abnormal relationship between the talus and os calcis. The patient is positioned supine, with the feet at the end of the operating table where the surgeon sits comfortably. Bilateral cases are treated in one session. I always start with a posteromedial release. It consists of the posterior release and a medial release of the subtalar joint, without sectioning the interosseous ligament between the talus and os calcis. The posterior tibial tendon is lengthened by a supramalleolar Z-plasty, the tendons of the long toe flexors are operated on only if they are very short. If the foot shows an obvious cavus and adduction deformity, a tenotomy of the abductor hallucis and sectioning of the plantar fascia is carried out. For the latter, usually an extra plantar incision is used to prevent injury to neurovascular structures. Sometimes a medial capsulotomy of the first tarsometatarsal joint is done. The most important step in the operation is the realignment of the subtalar complex. It requires a plantar, medial and dorsal capsulotomy of the talonavicular joint. Because the talus is displaced towards the medial malleolus, a careful, more distally orientated dissection of the joint by means of a pair of scissors is performed, to prevent cutting the talar neck. The spring ligament is cut and the navicular must be repositioned in front of the talar head. If, after this medial release, the talonavicular joint cannot be reduced completely and with ease, an extra incision is placed over the anterolateral aspect of the foot. The lateral part of the talocalcaneal joint is opened, as also is the calcaneocuboid joint. The tight capsule covering the talar head is removed until the cartilage of the talar head can be seen. Now the anterior part of the os calcis and the cuboid can derotate from under the talus and the talar head can be put into a proper position relative to the navicular and first ray of the foot. The reduction of the subtalar complex is maintained by percutaneous pinning of the talonavicular joint. The other joints are not pinned. After suturing of the Achilles tendon and the posterior tibial tendon the tourniquet is released. No vigorous attempt is made to achieve a completely dry wound, as there will always be some bleeding from joint capsules, and an excessive use of the electrocautery will probably do more harm. The wound is closed and an above-the-knee plaster is applied. It is important to avoid an extreme position of correction, because it will lead to too much tension on the skin edges. An extensive dissection of the tarsus is painful and requires adequate postoperative analgesia. A sacral anaesthetic block and a morphine infusion will keep the child and the parents comfortable in the days after the operation.

Postoperative management

After a simple posterior release, the sutures are removed after 2 weeks and a new cast is applied for a further 4 weeks. Then a simple below-the-knee splint, that will keep the foot in plantigrade position, is manufactured. Until the child starts walking, the splint has to be worn about 20 hours a day and thereafter only at night. I do not prescribe modified shoes for a child with well corrected feet.

After a more extensive release, the plaster is changed 4 days after the operation in the plaster-room without anesthesia and when the wound looks satisfactory the child is discharged from the hospital. Two weeks after the operation, the sutures are removed and the foot is put into a plaster again for another 6 weeks, with the foot in full correction. The K-wire is removed at the end of the immobilization period. Manufacture of the splint and its use are the same as after a posterior release. Usually, I continue the splinting at night until age 4 or 5. No special or modified shoes are prescribed after extensive release.

Treatment of relapse

Real recurrence of the deformity is rare. Most recurrences are, in fact, undercorrected deformities. Our hospital is a secondary referral center and therefore we see many children for secondary treatment. Close

to half of the cases referred have been operated on earlier. If an undercorrected clubfoot is seen, a precise assessment of the residual deformity is mandatory. Clinical and radiographic examinations, in which the lateral views are taken in maximum dorsiflexion and plantarflexion will give a detailed picture of the deformity, the mobility of the various joints, the sometimes abnormal shape of the bones of the foot and the probable overcorrection in other joints. Tarraf and Carroll (1992) have described extensively the residual deformity in the feet which were referred to them for secondary treatment. I have learnt much from their experience and highly recommend reading their study.

If in the first operation only a limited soft tissue release has been performed, a new but more extensive release may be done until age 4 or 5. The commonest problems are the insufficient release of the posterolateral tether and the undercorrection of the reduction of the subtalar complex. In these cases a calcaneocuboid resection and fusion (Evans 1961) is sometimes necessary to straighten the foot. I have no personal experience of the lengthening procedure of the medial ray described by Hofmann et al. (1984). A special problem in revision cases is the sometimes quite severe scarring and shortening of the skin and subcutaneous tissue on the posteromedial side of the foot and ankle. I often perform Z-, double Z- or W-plasties to close the skin without undue tension.

In children older than 6 years, it is hardly ever possible to create a plantigrade foot by soft tissue release only. Metatarsal osteotomies correct severe residual forefoot adduction. A triple fusion to modify a stiff varus hindfoot into a plantigrade stiff foot is mostly postponed until age 12 or 13. I have no personal experience of isolated valgus osteotomy of the os calcis (Dwyer 1963). Severe rotational deformity of the leg may require a corrective supramalleolar osteotomy. I did not need to use it in the treatment of idiopathic clubfeet.

A common feature of all revision surgery is a higher complication rate, like disturbed wound healing and infection, and that the result is usually less satisfactory than that of primary surgery. In reoperated clubfeet, the ultimate shape of the foot is often rather good, but the mobility of the ankle and tarsus remains severely restricted in almost all cases.

Evaluation and follow- up

As in many pediatric orthopedic problems, growth may have a positive or negative influence also on the ultimate outcome of clubfeet. It is therefore important to organize a follow-up at regular intervals, at least until maturity. During the period the foot is splinted during sleeping hours, the child is seen every 6 months to check the splint and the form and function of the foot. After the age of 4, after abandoning the splint, the child is seen every year and later on every 2 years.

With the treatment protocol I have described , we operate on 80% of the cases treated primarily by closed methods. This rate may seem high, but is based on the strict criteria we use for the evaluation of the closed treatment. A posterior or posterolateral release is done in about 40%, a posteromedial release in 30% and the remaining 30% need a complete peritalar release. The reoperation rate in the cases primarily treated in our center is less than 10%.

There are still some problems to be solved in the treatment of idiopathic clubfeet. Despite all efforts, it is impossible to cure the problem completely. Most well treated clubfeet show restricted motion of the ankle and tarsus. The feet which were very stiff from the start of treatment, can be brought into a plantigrade position relative to the leg and are usually painless during childhood and adolescence, but the function in adult life has yet to established. Another problem is the slender and straight shape of the calf which is often quite obvious in unilaterally affected girls. It is not rare to hear that this cosmetic feature is disturbing her the most! Extra exercises to build muscle mass do not help.

A discrepancy in foot length may occur even in bilaterally affected cases. It hardly ever exceeds 3 shoe sizes and may be managed by modification of the shoes. A discrepancy in leg length is usually less than 2 centimeters and does not require any treatment.

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