■ CHILDREN'S ORTHOPAEDICS

The management of the orthopaedic sequelae of meningococcal septicaemia

PATIENTS TREATED TO SKELETAL MATURITY

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Septicaemia resulting from meningococcal infection is a devastating illness affecting children. Those who survive can develop late orthopaedic sequelae from growth plate arrests, with resultant complex deformities. Our aim in this study was to review the case histories of a series of patients with late orthopaedic sequelae, all treated by the senior author (CFB). We also describe a treatment strategy to address the multiple deformities that may occur in these patients.

Between 1997 and 2009, ten patients (seven girls and three boys) were treated for late orthopaedic sequelae following meningococcal septicaemia. All had involvement of the lower limbs, and one also had involvement of the upper limbs. Each patient had a median of three operations (one to nine). Methods of treatment included a combination of angular deformity correction, limb lengthening and epiphysiodesis. All patients were skeletally mature at the final follow-up. One patient with bilateral below-knee amputations had satisfactory correction of her right amputation stump deformity, and has complete ablation of both her proximal tibial growth plates. In eight patients length discrepancy in the lower limb was corrected to within 1 cm, with normalisation of the mechanical axis of the lower limb.

Meningococcal septicaemia can lead to late orthopaedic sequelae due to growth plate arrests. Central growth plate arrests lead to limb-length discrepancy and the need for lengthening procedures, and peripheral growth plate arrests lead to angular deformities requiring corrective osteotomies and ablation of the damaged physis. In addition, limb amputations may be necessary and there may be altered growth of the stump requiring further surgery. Long-term follow-up of these patients is essential to recognise and treat any recurrence of deformity.

Meningococcal infection presenting as bacterial meningitis or meningococcal septicaemia is a leading cause of morbidity and mortality in early childhood. The causative organism is a Gram-negative diplococcus, *Neisseria meningitidis*, whose endotoxin triggers a complex cascade of events including complement activation, the release of interleukin and tumour necrosis factor, diffuse vasculitis, disseminated intravascular coagulation, septic shock and multi-organ failure. Although the introduction of vaccines to control Hib, serogroup C meningococcus and some types of pneumococcus has dramatically changed the epidemiology of the disease, infections caused by serogroup B continue to predominate in the United Kingdom and Europe.¹ Children who survive severe infection may have serious long-term morbidities, including hearing loss, orthopaedic, dermatological, renal and neurological, developmental complications, and psychosocial problems.

In meningococcal septicaemia, disseminated intravascular coagulation may cause clinically obvious haemorrhagic skin necrosis. Occult damage to bones and growth plates may also occur. An initial vascular occlusive process leads to an inflammatory process that damages the bone and cartilage of the metaphysis, physis and epiphysis.² Damage to the growth plate can present some years after the acute illness as either a limb-length discrepancy (with a central growth plate arrest) or a combination of limb discrepancy and angular deformity (with a peripheral growth arrest).

A better understanding of the initial pathophysiology of the disease and the resultant limb deformities may help the orthopaedic surgeon to manage these patients. There have been several reports in the literature of the orthopaedic sequelae of meningococcal disease,³⁻¹⁰ but little guidance on the management of these patients over the course of their skeletal growth. The aim of this retrospective

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Fig. 1

Bar chart showing the distribution of clinical presentation following meningococcal septicaemia (LLD, limb-length discrepancy).

study was to review the senior author's (CFB) experience of treating patients referred with sequelae of meningococcal septicaemia with follow-up to skeletal maturity.

Patients and Methods

Between 1997 and 2009, ten patients with late orthopaedic sequelae after meningococcal septicaemia were treated by the senior author (CFB). They were managed at two tertiary referral hospitals in the United Kingdom. Four of these patients have been previously reported in a multicentre study by Belthur, Bradish and Gibbons¹¹ which looked at the management of 24 patients with late sequelae following meningococcal septicaemia. We have incorporated these four patients into our series with further information on their treatment and follow-up to skeletal maturity.

The definitive diagnosis of meningococcal infection was established either by a positive blood culture, cerebrospinal fluid or skin lesion, or by counterimmune electrophoresis and latex particle agglutination tests that detect capsular antigen. There were seven girls and three boys with a median age at referral of 8.3 years (3 to 12) and each had a median of three operations (one to nine). The mean follow-up was 66 months (24 to 126). For the purpose of this study a limb-lengthening procedure with application and subsequent removal of an external fixator, combined with an epiphysiodesis if needed, was considered as one operation. The medical records and radiographs were reviewed retrospectively. Data were collated and analysed on an Excel spreadsheet (Microsoft Corp., Seattle, Washington).

Results

The most common presentations were limb-length discrepancy and angular deformity at the knee and/or ankle (Fig. 1, Table I). One patient had bilateral wrist deformities in addition to a lower limb-length discrepancy, and one presented with an angular deformity in her right below-knee amputation stump. In total, these ten patients had growth arrests involving 27 sites, the most common of which were the proximal and distal tibia. Eight patients had

multiple growth plate involvement, ranging from two to five growth plates. Growth arrests occurring around the knee resulted in varus deformity with or without recurvatum in six of eight patients (Fig. 2). One patient (case 1) with involvement of the distal femoral and proximal tibial growth plates had a deformity of the lower limb without malalignment of the knee. Another (case 8) with involvement of the left distal femoral growth plate had a deformity of the lower limb without malalignment of the knee.

All of the six patients with an angular deformity around the knee had angular correction with or without lengthening using either an Ilizarov frame or a Taylor Spatial Frame (Smith & Nephew Richards, Memphis, Tennessee). One patient (case 3) who had an epiphysiodesis of the proximal tibial growth plate at the index operation had angular correction and lengthening without recurrence of deformity. One patient (case 9) had an angular correction and lengthening with a transphyseal osteotomy of her below-knee amputation stump without recurrence of deformity.

Growth plate arrests of the distal tibia occurred in six patients, invariably resulting in varus deformity (Fig. 3). The distal fibular growth plate tends to be spared from growth plate arrest and so becomes relatively overgrown in relation to the tibia. The relationship between the distal tibia and fibula must be considered in any corrective procedure. Four patients (cases 2, 5, 7 and 8) had an acute correction with a transphyseal distal tibial osteotomy held with screws (Fig. 4). One patient (case 6) had tibial lengthening and varus ankle correction with a distal tibial corticotomy using a Taylor Spatial Frame, and once the distal tibiofibular joint was level a fibular wire was added and a fibular osteotomy performed. One patient (case 10) had a proximal tibial corticotomy and lengthening with a Taylor Spatial Frame together with acute correction of the varus deformity of the ankle with a supra-malleolar dome osteotomy.

One patient (case 1) had growth plate arrests of both her radii, resulting in deformity of her forearms and radial deviation of her wrists. This was treated by lengthening the radius using a monolateral external fixator. After removal of the external fixator from her right and left forearms at one and three years, respectively, she has full range of movement of both wrists and restoration of the normal alignment of both distal radioulnar joints.

One patient with bilateral below-knee amputations had satisfactory correction of a deformity of the right stump and ablation of both proximal tibial growth plates. She now uses her prosthesis without problems.

Eight patients have had their leg-length discrepancy equalised to within 1 cm, with restoration of the normal mechanical axis of the lower limbs. One patient (case 3) is significantly short (7 cm), with a fixed flexion deformity, but has declined further surgery.

Complications. Three patients (cases 2, 4 and 7) who had epiphyseal stapling of the lateral proximal tibial growth plate as an adjunct to correction of an angular deformity had recurrence of the original deformity (Table II). Subsequent

Table I. Clinical details of the 12 patients

Case	Gender	Age at referral	Reason for referral*	Growth arrest number/sites†	Number of operations
1	F	4 yr/8 m	LLD (short rt LL) & both forearm radii	4/DF, PT, DR×2	5
2	F	3 yr/2 m	LLD varus both knees, varus rt ankle	5/PT×2, DT×2, DF	9
3	M	10 yr	LLD (short rt LL), rt knee recurvatum & varus	2/DF, PT	3
4	M	3 yr/6 m	LLD varus both knees	4/DF×2, PT×2	5
5	M	7 yr/8 m	LLD (short rt LL), equinus rt ankle	1/DT	5
6	F	12 yr	LLD varus rt ankle	1/DT	2
7	F	9 yr	LLD varus both knees, varus rt ankle	4/PT×2, DT×2	2
8	F	7 yr	LLD short lt femur, varus both ankles	3/DF, DT×2	2
9	F	11 yr	Rt below-knee amputation stump deformity	1/PT	1
10	F	10 yr	LLD (short lt LL), varus lt ankle	2/PT, DT	2

* LLD, leg-length discrepancy; rt, right; lt, left; LL, lower limb

† DF, distal femur; PT, proximal tibia; DR, distal radius; DT, distal tibia

Fig. 2

Radiograph showing growth plate arrests around the knee resulting in significant varus deformities (treated with bilateral tibial osteotomies prior to referral).

Fig. 3

Radiograph showing growth plate arrests of the right distal tibia resulting in varus deformity at the ankle.

operations incorporated an epiphysiodesis of the proximal tibial growth plate, with no further recurrence of the angular deformity.

One patient (case 3) developed posterior subluxation of his right knee following femoral angular correction and lengthening. This was a result of an anterior slope to his tibia causing a recurvatum deformity and predisposing to posterior subluxation of the knee. The lengthening procedure was stopped and his knee reduced spontaneously. He has a residual 10° fixed flexion deformity of the knee and a significant limb-length discrepancy of 7 cm. He has declined further surgery.

One patient developed a deep infection at the proximal tibial pin sites. This did not respond to antibiotics and required formal incision and debridement. The treatment was successful and two years later she has had no further symptoms or signs of infection.

Discussion

Meningococcal septicaemia remains a major cause of morbidity and mortality in most countries. Patients who

survive often have serious long-term orthopaedic sequelae. Buysse et al¹² found a significant correlation between the severity of the disease and major skin and orthopaedic sequelae. Bache and Torode¹³ described 16 patients with skeletal sequelae; in whom there were 41 cases of physal arrest. They also found that amputation was invariably associated with subsequent proximal growth plate disturbances. Belthur et al¹¹ similarly found 49 growth plate arrests in 23 patients. Such growth arrests can result in complex deformities and present a huge challenge to the treating orthopaedic surgeon.

There are two main theories as to the pathogenesis of these growth plate arrests. The first postulates an ischaemic insult secondary to the effects of the *N. meningitidis* endotoxin, which initiates a diffuse vasculitis and disseminated intravascular coagulation. There is an acute vascular occlusive process that renders bone ischaemic, and later a discrete inflammatory response that further contributes to this ischaemia.^{2,12,14,15} A second theory was suggested by Watson and Ashworth,⁷ who postulated that, as with burn scars, areas of necrotic skin could act as a tether, producing

Fig. 4a

Fig. 4b

Radiographs showing acute correction of a varus deformity at the ankle with a transphyseal distal tibial osteotomy (a) held with two screws (b).

a type 6 growth plate injury involving only the periphery of the plate and particularly the ring of Ranvier. However, Belthur et al¹¹ observed that areas of skin necrosis did not necessarily overlie areas of future growth plate damage, thereby lending more weight to the theory of ischaemic damage.

There is relatively little guidance for the orthopaedic surgeon managing these conditions, and we therefore reviewed this experience of treating these patients to skeletal maturity and present some principles of management.

A thorough assessment is crucial with a full clinical examination and complete radiological investigations. Clinical examination, radiographs and CT scans are used to assess the deformity and limb-length discrepancy, and to identify which growth plates are affected and to what extent. It is important to assess the function of the joints affected, particularly the knee and ankle joints, which often present with severe varus deformities. Prediction of final limb-length discrepancy is also important in order to anticipate the need for further lengthening. At first presentation this can be undertaken in a simple manner by using the arithmetic method, with the distal femur contributing about 9 mm of growth each year, the proximal tibia about 6 mm and the distal tibia about 5 mm.^{16,17} Later, with serial measurements, the estimate can be made more accurately using the multiplier method of Paley et al.¹⁸

In a study looking at the management of these deformities carried out at six tertiary centres, Belthur et al¹¹ observed that patients with growth arrests around the knee who underwent realignment procedures (either with an osteotomy or by hemicallotasis) without an additional epiphysiodesis had a high rate of recurrence of deformity. They found that 15 of 16 patients treated for an angular deformity around the knee without an epiphysiodesis had recurrence of deformity.

In our series, angular deformities were corrected using either hemicallotasis with a circular external fixator or by

conventional closing or opening wedge osteotomy. Epiphysal stapling in an attempt to correct a deformity was invariably unsuccessful. One patient with a distal femoral growth plate tether had an attempted epiphysiodesis with a fat pad graft that failed, resulting in a recurrent femoral varus deformity. The extent of the physeal damage was probably underestimated, and for this reason surgeons should be cautious about proceeding to epiphysiodesis.

All patients who had an angular correction and lengthening combined with a percutaneous epiphysiodesis had no recurrence of angular deformity. The role of epiphysiodesis in preventing recurrent deformity has also recently been highlighted by Nectoux et al.^{9,10}

Varus deformity around the knee as a result of meningococcal infection is similar to that caused by Blount's disease, with a variable extent of medial tibial plateau involvement. Hemiplateau elevation and bone graft may be necessary to restore joint congruity. Complex deformities are seen in the leg as a consequence of a combination of proximal and distal tibial arrests together with relative overgrowth of the fibula, whose growth plates are typically spared in this condition. The tibia may simply be short, or short and angulated, and the site of deformity may be proximal, distal or both, depending on the growth plates involved. Typically a varus knee deformity is seen, but if more of the anterior growth plate is involved a recurvatum deformity will develop.

When the deformity is purely in the proximal tibia, lengthening together with any necessary angular correction is undertaken, and an epiphysiodesis of the proximal tibia and fibula is performed to prevent recurrence of angular deformity. With a distal tibial growth arrest the resultant deformity is usually a varus angulation with overgrowth of the fibula. In our series, four patients had a distal tibial transphyseal osteotomy which ablated the growth plate and enabled an acute correction of the tibial varus deformity.¹⁹ This procedure is appropriate if the normal distal

Table II. Details of the operations and outcomes

Case	Procedures in chronological order*	Complications	Final outcome†
1	Rt tibial lengthening with CEF Rt femoral lengthening with CEF Bilateral radial lengthening with monotube external fixator		No LLD, good correction both forearms
2	Bilateral tibial angular correction with CEF Rt PT lateral epiphyseal stapling + transphyseal DT osteotomy Lt PT lateral epiphyseal stapling Rt tibial angular correction + PT epiphysiodesis (drilling) Lt femoral DF epiphysiolysis + fat pad graft Lt femoral angular correction with CEF Lt medial PT hemiplateau elevation with CEF Lt femoral angular correction with CEF	Recurrence of bilateral knee varus deformity following epiphyseal stapling Recurrence of Lt knee varus deformity following epiphysiolysis	No LLD, normal alignment
3	Rt tibial angular correction + lengthening with CEF + PT epiphysiodesis (drilling) Rt femoral angular correction + lengthening with CEF		LLD (7 cm short rt leg), normal alignment, does not want further surgery
4	Rt femoral corrective osteotomy Rt tibial angular correction + lengthening with CEF Lt tibial angular correction with CEF Lt femoral corrective osteotomy Rt tibial angular correction + PT epiphysiodesis (drilling)	Recurrence of bilateral knee deformity	Mild LLD (1 cm short rt leg), normal alignment
5	Rt tibial lengthening + DT osteotomy + ankle arthrodesis with CEF Rt tibial osteotomy + plating following traumatic fracture through regenerate with resulting valgus malalignment Rt PT medial hemiepiphysiodesis (8 plate) for valgus malalignment Bilateral PT medial hemiepiphysiodesis (8 plate)	Recurrence of valgus deformity	No LLD, normal alignment
6	Rt tibial angular correction with CEF Adjustment of frame with fibular wire + osteotomy		No LLD, normal alignment
7	Bilateral lateral PT epiphyseal stapling Rt DT transphyseal osteotomy Bilateral PT epiphysiodesis + angular correction with CEF	Recurrence of deformity	No LLD, normal alignment
8	Lt femoral lengthening with CEF Bilateral DT transphyseal osteotomy		No LLD, normal alignment
9	Rt BKA stump angular correction + lengthening with CEF		No prosthetic problems
10	Lt tibial angular correction + lengthening with CEF + Lt DT supramalleolar osteotomy		No LLD, normal alignment

* Rt, right; Lt, left; CEF, circular external frame; DF, distal femur; PT, proximal tibia; DT, distal tibia; BKA, below-knee amputation

† LLD, leg-length discrepancy

tibiofibular relationship can be restored by a simple angular correction of the tibia. It must in addition be combined with a distal fibular epiphysiodesis to prevent recurrence. One patient had a supramalleolar dome osteotomy to achieve an acute correction. This has the advantage of respecting the centre of rotation of angulation in correction of the deformity, and must be combined with a distal

fibular epiphysiodesis. If the distal tibia is short in relation to the fibula, then tibial lengthening will be required. Distal tibial lengthening/angular correction to restore the tibiofibular relationship can be undertaken through a supramalleolar corticotomy. If overall lengthening of the tibia and angular correction is required because of a limb-length discrepancy, then differential tibial lengthening can be

performed with either a proximal tibial corticotomy or a combination of proximal and distal tibial corticotomies. A circular frame is applied to the tibia and the tibial corticotomies performed. The fibula is initially left intact and secured to the frame with a single proximal wire. This enables lengthening of the tibia alone. When the tibiofibular disproportion has been corrected the fibula is divided, a distal fibular wire inserted, and lengthening of both tibia and fibula continued until the desired length has been achieved.

There was only one case in this series with clinically significant forearm deformity, involving shortening of the radius. A slightly different strategy is recommended in the forearm. If there is cosmetic shortening (> 5 cm) then lengthening of both forearm bones will be required. If there is shortening of one bone alone causing radial or ulnar deviation of the wrist, then the involved bone alone will require lengthening/angular correction. When the normal radio-ulnar alignment has been restored an epiphysiodesis of both bones can be performed to prevent recurrence. In young patients this would create an unacceptable amount of forearm shortening; in the senior author's opinion, when there is a recurrent deformity it is preferable to repeat the lengthening procedure on the involved bone.

In conclusion, the surgical approach to correction of deformity depends on the extent and site of damage to the growth plate. If there is central growth arrest, the aim is to achieve limb length equality by lengthening. If there is a peripheral growth plate arrest, angular correction is also necessary, and we recommend ablation of the growth plate in the lower limb to prevent recurrent angular deformity. In the lower limb angular correction can be either acute, using a dome or transphyseal osteotomy, or gradual using a circular external frame. Follow-up to skeletal maturity is essential in order to recognise and treat any recurrence. The overall goal must be to achieve limb-length equality and normalisation of the mechanical axis in the lower limb with minimal loss of stature. The surgeon, the patient and carers must be prepared for repeated bone-lengthening surgery.

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