THE KLIPPEL-TRENAUNAY SYNDROME

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The Klippel-Trenaunay Syndrome is a triad of congenital anomalies characterised by a vascular naevus, varicose veins and hypertrophy of soft tissue and bone. A number of patients affected with this rare syndrome need amputation. In this paper the systemic problems, stump complications and prosthetic difficulties of four amputees with Klippel-Trenaunay syndrome are outlined. The period of follow-up ranged from 10 to 24 years after amputation.

Two French physicians, Klippel and Trenaunay (1900), described a clinical syndrome with three major features: 1) haemangioma; 2) hypertrophy of soft tissue and bone overgrowth; and 3) varicose veins. The varices tend to be located along the lateral aspect of the limb with the so-called 'lumbar-to-foot' pattern. They are often associated with suprapubic and perineal varices of varying severity, and Baskerville, Ackroyd and Browse (1985) reported a high incidence of thrombophlebitis and thrombo-embolic episodes. Limb length discrepancy is common, and gross gigantism with grotesque deformities and arteriovenous abnormalities tend to produce severe cosmetic and functional disabilities; because of its weight the limb is dragged along, often in marked external rotation. In a series reported from the Mayo clinic (Gloviczki et al 1983), 95% of cases were of the lower limb and 85% were unilateral.

CASE REPORTS

Case 1. In 1978 a five-month-old baby with a giant haemolymphangiomatous malformation underwent a left through-knee disarticulation. Five years later a lymphangioma involving both buttocks and the perineum was excised, and in 1988 a pelvic abscess was drained. The left thigh is longer than the right (Fig. 1) and he is currently using a modular prosthesis with an energy-

storing foot mechanism. The fluctuation of stump volume precludes him from using a total contact self-suspending prosthesis.

Case 2. In 1979 a 14-year-old boy with a giant haemolymphangioma had a left below-knee amputation; this was later revised to mid-thigh level because the wound broke down. Four to five years after the amputation he

Fig. 1

Eleven years after amputation the left through-knee stump is longer than the right thigh.

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developed verrucous lesions and lymphatic seepage from the distal end of the stump. The lymphorrhoea became profuse and warranted abdominal ligation of lymphatics. He is, at present, using a modular prosthesis with a pelvic belt suspension, but frequent stump changes interfere with its use.

Case 3. In 1969 a 10-year-old boy underwent a right above-knee amputation for haemolymphangiomatous malformation and gross limb hypertrophy. Five years later he had multiple gluteal and stump swellings excised. He has had systemic manifestations of the syndrome with pyrexia of unknown origin. The thermoplastic socket on his modular prosthesis has a quadrilateral brim. He has, in the past, tried an ISNY (Iceland-Sweden-New York) type of socket, but could not continue with this because of changes in the stump volume.

Case 4. In 1958 a five-year-old boy had haemolymphangiomatous swellings of the left lower thigh and calf excised. This was followed by a fixed flexion deformity of 90° at the knee and recurrent calf swellings. Tethering of the tibial nerve in the vascular tissue resulted in a painful leg. In 1965 he had a left mid-thigh amputation and subsequently, vascular lesions of the gluteal and trochanteric regions have been excised. A CT scan showed other vascular malformations in the pelvic region, but these were left untreated. He is able to use a modular prosthesis made of carbon fibre. The socket is of the self-suspending type and he overcomes the stump-socket volume mismatch by using two or three leather linings.

DISCUSSION

In the Mayo Clinic series (Gloviczki et al 1983) 40 of the patients (67.5%) were treated non-operatively. Indications for surgery include functional disability, cardiac failure, and cosmetic reasons. Control of foot size can be achieved by metatarsal epiphysiodesis combined with ray amputation, and limb length discrepancy by epiphy-

siodesis of the lower femur and upper tibia and fibula. Amputation may be needed, but it is not easy to decide on the level. In children, a through-knee disarticulation is preferred to a mid-thigh amputation because the growth plate of the distal femur is preserved, the operation is less traumatic (which is important in the presence of excessive vascular tissue), and also because it gives an end-bearing stump with good rotational stability.

Mid-thigh amputations do not achieve total clearance of the vascular malformation, as gluteal and pelvic areas are often involved. Moreover, these amputations necessitate weight transmission through the ischium and any gluteal swellings or scars may present problems. Another difficulty is that the volume of the stump may vary with changes in the vascular content; flexible and semi-flexible liners have been used in rigid socket frames to accommodate for these volume changes. A total surface bearing type of self-suspending socket to give uniform pressure on the soft tissues of the stump is another option. The recently introduced above-knee socket with ischial containment – so called CAT-CAM (contoured adducted trochanteric, controlled alignment method, Schuch 1988), has a high gluteal brim which was not acceptable to our patients. Clearly, a pre-amputation consultation with a prosthetic clinician is advisable.

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