CONGENITAL ARTERIOVENOUS FISTULAE OF THE EXTREMITIES

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FIG. 1.—Ventral view of the left fore-limb of a 12 mm. pig embryo. Of the segmental arteries which initially supply the fore-limb, only the stem of the seventh remains to form the subclavian artery. The main vessels in the limb are becoming differentiated from a diffuse capillary network as retiform channels. (From H. H. Woollard.)

Patients suffering from this curious vascular malformation provide an interesting study for the vascular surgeon, but they may present in the practice of many branches of medicine. Persistent ulceration may bring them to varicose vein or plastic surgery clinics; hypertrophy of bones or soft tissues may be responsible for attendance at paediatric or orthopaedic clinics and finally the remote effects on the heart sometimes cause the patient to be referred to cardiological clinics.

Embryology

Real understanding of the nature of this condition followed the researches of Sabin (1917-18) and Woollard (1922). Their studies of the development of blood vessels in the limb are of fundamental importance, since they afford an acceptable explanation of the origin of these multiple abnormal connections between arteries and veins. The stages in development were studied in chick

and in pig embryos and the results may be summarized as follow:

1. The angioblastic stage.
2. The stage of the capillary network. It will be remembered that both arteries and veins have a similar origin in the capillary plexus.
3. The retiform stage, which is characterized by the presence of enlarged tubes tending to coalesce.
4. Finally, the formation of a definite stem.

Fig. 1 illustrates the transition from the capillary plexus to the retiform stage by the persistence of the more fortuitously placed capillaries and the disappearance of their less fortunate neighbours. It will be appreciated that, should these capillaries not disappear, then the condition of congenital arteriovenous fistulae will occur. The multiplicity of the abnormal connections, as seen in the fully developed condition, is also explained. Workers, such as Rienhoff (1924), Reid (1925), Horton

FIG. 2.—Note elongation of affected left middle finger and distension of superficial veins.

FIG. 3.—Note swelling of the left hypothenar eminence and dilatation of superficial veins in left forearm.

FIG. 4.—Arteriogram of patient shown in Fig. 3. Note tortuous and dilated ulnar artery and abnormal vessels around base of fifth metacarpal.

(1932) and de Takats (1932), were quick to realize the implications of the studies of Sabin and of Woollard and to apply them to the clinical field.

Applied Physiology

It is convenient at this point to recall the possible effects of arteriovenous fistulae. These were classified by Pemberton and Saint (1928) as local, regional and systemic.

Local Effects. There is dilatation of the super-

ficial veins and, as a result of increased venous pressure, hypertrophy of the vein wall results. The arteries proximal to the lesion also show dilatation.

Regional Effects. Increase in growth in length and in girth of the bones is seen if the fistulae are open before the epiphyses unite. Frequently the soft tissues also show hypertrophy. There is an increase in surface temperature. Nutritional lesions may be present and may be limited to ulceration, but gangrene may occur.

Systemic Effects. According to Holman (1937), the presence of arteriovenous fistulae provides two circuits through which blood may flow—through the normal capillary bed, or through the fistulae. The reduced peripheral resistance in the fistulae attracts blood to this circuit and causes a reduction in blood pressure, especially the diastolic, in the first circuit. Compensatory mechanisms are evolved, which are designed to maintain the blood pressure in the capillary bed at an adequate level. These include an increase in the pulse rate and an increased circulating blood volume. In certain patients cardiac enlargement occurs. An interesting circulatory phenomenon found in certain patients with arteriovenous fistulae is the *bradycardiac reaction*. This term is applied to the slowing of the pulse which follows occlusion of the main artery of the affected limb. A positive bradycardiac reaction is evidence that a considerable volume of blood is being short circuited.

Classification of Cases

Analysis of a large series of patients with vascular malformations in the extremities (Robertson, 1953-56) has shown that the lesions may be either *localized* or else *diffuse*.

Localized Lesions

Localized congenital arteriovenous fistulae are often situated in the peripheral part of the extremity and are usually present for a number of years before symptoms develop.

The presenting symptom is frequently ulceration, but recurrent haemorrhage may also necessitate consultation, as may the presence of a pulsatile swelling. The characteristic physical signs of an arteriovenous communication are present in most cases, i.e. local increase in surface temperature, dilatation of the proximal arteries and superficial veins and a continuous murmur is audible in the region of the fistulae. Gigantism of the part is not striking, but in most patients there is a slight increase in the length of the bones in the region of the fistulae.

In these localized or peripheral lesions arteriography is the most useful special investigation. It permits accurate definition of the extent of the

FIG. 5.—Arteriogram showing extensive lesion involving distal part of forearm and the hand.

lesion and this is of particular importance if surgical excision or amputation is contemplated. The blood flow in these cases is extremely rapid and the best results are obtained by serial angiography. Complicated apparatus is not necessary, a manually operated cassette tunnel being all that is required.

Localized lesions in the upper extremity may be restricted to a digit (Fig. 2). Less commonly the hand is also involved (Fig. 3). Arteriography (Fig. 4) shows the abnormal vessels and the characteristic angiographic features associated with congenital arteriovenous fistulae. These are pooling of the dye in the region of the fistulae, dilatation of the proximal arteries and deprivation of blood supply to the peripheral tissues. In two patients seen by the writer the deprivation of blood supply to the periphery was so extreme as to lead to gangrene.

In the lower limb the lesions may be confused with primary varicose veins. The affected limb will, however, be noted to be slightly longer than the normal one; there will be an increase in surface temperature and a bruit will be audible.

Cardiovascular Effects

It is very rare for cardiac enlargement to be present in association with localized congenital arteriovenous fistulae. Rarely, the bradycardiac reaction is positive.

Prognosis

Blood flow through the abnormal vessels tends to increase with time and ulceration or gangrene is often the end result. However, many years may pass before these complications occur and unless the lesion is causing pain or is extremely circumscribed surgical treatment should not be undertaken in the absence of complications.

The indications for treatment may be ulceration, gangrene, recurrent haemorrhage and, occasionally, pain.

Treatment

The ideal, which experience shows is rarely possible, is complete excision of the lesion. It was possible in four patients in a series of 14 personally studied.

Extensive lesions in the digits are best treated by amputation of the digit. In the hand lesions which are too extensive to permit complete removal they may be treated by subtotal excision, combined if necessary with partial amputation of the hand. Fig. 5 shows the arteriogram of a patient with a gross lesion in the wrist and hand. Arteries proximal to the lesion had been ligated 20 years previously and about the same time gangrene of the fifth finger necessitated amputation of this digit. An ischaemic ulcer on the dorsum of the hand necessitated further treatment in 1950. The

FIG. 6.—Result after partial amputation of the hand in patient whose arteriogram is shown in Fig. 5. The hand remains soundly healed six years after operation.

ulcer was acutely painful and flexion contractures were present in the third and fourth fingers. Subtotal excision of the abnormal vessels was combined with partial amputation of the hand and the remnant remains soundly healed six years later, the patient continuing to work as a clerk (Fig. 6).

Extensive lesions in the lower extremity in female patients are best treated by subtotal excision, in the first instance, if complete removal is not possible, for preservation of the extremity is all important and may be achieved over a period of years. In male patients with gross lesions a well-timed below knee amputation may reduce invalidism and be in the patient's best interests.

Ligation of the proximal main arteries is contraindicated. At the best, the results of such ligations are only temporary; at the worst, gangrene may well be the result.

Diffuse Congenital Arteriovenous Fistulae

When the abnormal connections between arteries and veins are widespread throughout the extremity gigantism of the limb is the rule. The typical clinical picture is that of a giant limb which is usually noted early in life (Fig. 7). The increase in the length of the limb can be clearly seen and usually it affects all bones in the limb, including those of the hand and foot. Superficial angiomas

are also obvious and were present in 20 of the 28 patients studied (Robertson, 1956). An increased surface temperature and varicose veins are other characteristic features. It should be emphasized that bruits and thrills are unusual in these giant limbs. When present these signs indicate a gross diversion of blood flow and suggest a poor prognosis for the limb in these patients.

Although the increase in girth of the limb is usually commensurate with the lengthening, sometimes it is so gross as to resemble lymphoedema, which may, indeed, be present in addition to the vascular anomaly. Patients with such soft tissue hypertrophy have an increased liability to recurrent cellulitis due to a streptococcal infection. Originally noted in patients with lymphoedema, this infection is sometimes known as elephantiasis fever.

Other complications which have been encountered are recurrent ulceration and recurrent haemarthrosis of the knee. The latter complication is the result of involvement of the synovial membrane by haemangiomatous tissue.

Cardiovascular Effects

In patients with giant limbs the bradycardiac reaction is not infrequently positive and cardiac enlargement is not uncommon.

FIG. 7.—Note elongation of left lower extremity and presence of superficial angiomata.

FIG. 8.—Lymphoedema associated with diffuse congenital arteriovenous fistulae. Note increase in length and girth of limb and presence of superficial angiomata.

Special Investigations

Plethysmography. This is probably the most useful diagnostic investigation in patients with giant limbs. Even when little clinical evidence exists of an arteriovenous shunt blood flow studies regularly demonstrate an increased flow in the affected limb.

Arteriography. The results of arteriography are disappointing, probably because fistulae are often present in relation to the smallest vessels only. Thus in only five of the 20 patients studied in a recent series (Robertson, 1956) were the abnormal vessels demonstrated.

Prognosis

If there is clinical evidence of a free diversion of the blood flow from arteries to veins, then the prognosis for the limb is poor. Thus, if the limb in childhood is large and hot and the bradycardiac reaction positive, the tendency is for complications to occur. These may be local, such as ulceration or cellulitis, or systemic, such as cardiac enlargement. On the other hand, if lengthening is more marked than soft tissue hypertrophy, and if the vascular effects are not prominent, then the prognosis for the limb is quite good.

Indications for Treatment

These include ulceration, recurrent cellulitis recurrent haemarthrosis, increased length or girth of the limb and, finally, cardiovascular complications.

Treatment

Since the fistulae, if they can be demonstrated, are usually multiple, it is not possible to carry out excision. Fortunately, in many patients the presence of the fistulae does not constitute a danger either to life or limb and symptomatic treatment may therefore be worth while.

The disparity in length of the bones of the lower extremity may be considerable and a compensatory scoliosis inevitable. This can be corrected by building up the shoe on the unaffected side and in many patients this is all that is necessary. If the disproportion in length is considerable and the prognosis for the limb favourable from the vascular point of view, leg equalization by means of epiphyseal stapling is justified.

When the girth of the limb is such as to resemble lymphoedema, and providing the prognosis for the limb is good, excision of the excessive superficial tissues and replacement of the skin on to the muscles is indicated.

Recurrent haemarthrosis, due to involvement of the synovial membrane of the knee joint by haemangiomatic tissue, may necessitate synovectomy and a good result may be expected, as judged by freedom from recurrence and function of the joint.

Amputation may be required in order to rid the patient of a heavy and grotesque limb which is affected both by intractable ulceration and repeated attacks of cellulitis (Fig. 8).

Cardiac decompensation constitutes an absolute indication for removal of the limb, but it must be remembered that cardiac enlargement *per se* is not necessarily an urgent indication for surgery. Slight degrees of enlargement, detected by X-rays, may be well tolerated by children, but a close follow-up should be instituted so that progressive deterioration may be treated by amputation of the affected extremity.

Summary

The origin of congenital arteriovenous fistulae of the extremities has been related to the development of blood vessels in the limbs.

The lesions may present as a localized or diffuse form.

The clinical features and treatment of the two types have been described and are summarized in the accompanying table.

Presenting Symptom	LOCALISED LESIONS	DIFFUSE LESIONS
	Ulceration Haemorrhage Localised swelling	Giant Limb
Obvious arteriovenous communication	Frequent	Infrequent
Superficial angiomas	Rare	Frequent
Lengthening of bones	Slight to moderate (in region of fistulae)	Marked (all bones in extremity)
Positive bradycardiac reaction	Occasionally	Not infrequent
Cardiac enlargement	Rare	Not infrequent
Arteriography	Always positive	Rarely positive
Surgical excision	Occasionally possible	Not indicated

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