Aspects of Plastic Surgery

Other Congenital Abnormalities

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Summary

The plastic surgeon is not a miracle worker, as so many of his patients believe. Nevertheless, he can do much to minimize the functional and cosmetic effect of many congenital deformities. If a moral can be drawn from this article it must be that the plastic surgeon should be given an early opportunity to see and assess the patients described here, if only to ease the anxiety in the minds of their parents by appropriate reassurance and discussion.

The very title of this article implies a hotch-potch: a necessarily brief description of the cases of congenital abnormality referred to the average plastic surgeon, and their treatment. I will try to neglect the rare deformities seen by plastic surgeons once a decade—and by general practitioners rarely once in a lifetime of practice—in favour of the more common but frequently as challenging problems that are brought to the out-patients department.

Of course, there is no such animal as “the average plastic surgeon.” Not only will his own training and interest influence the type of case he deals with, but so will the ability and interest of the surgeons in other specialties with whom he works. In various parts of the country many of the conditions about to be described are treated by other specialists and are by no means necessarily the prerogative of the plastic surgeon.

The initial Consultation

It must be realized that not only do the parents feel natural anxiety about their child: they also feel vaguely guilty that their product is imperfect and, furthermore, usually want to insure themselves against later charges from their offspring that they have not turned every available stone to procure the best end product.

National Health Service clinics are crowded, and consultants are busy. Nevertheless, every extra minute spent with the parents discussing the future problems of the child's treatment is an extremely valuable investment. The probable course of events must be described, with the timing of the various operations clearly stated, together with what results are to be expected from each operation. In the older child school examinations must be considered and the decision taken whether term time or holiday time is more easily sacrificed. With any patient the initial consultation is important in establishing a working rapport; with the parents of the deformed child it is doubly so.

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Misfortunes rarely come singly. Congenital deformities may be multiple. Remember to look for the concealed defect and not be blinded by the obvious superficial deformity.

Skin Tumours

Vascular abnormalities and pigmented naevi are common, and a frequent source of concern to the parents.

STRAWBERRY NAEVUS

This mixed capillary and cavernous haemangioma (fig. 1) is relatively easy to diagnose and even easier to treat. It forms a purplish or reddish compressible swelling anywhere in the body, but is more frequently seen on the trunk or close to it or on the head and neck. The key to diagnosis is in the history. It is never present at birth, but always develops a few days after. The lesion grows gradually during the first nine to twelve months of infant life, and then starts to regress. This regression is often heralded by the appearance of spreading white patches on the purple surface. At around 6 years of age the lesion may have vanished entirely, or may be represented merely by a loose skin tag, which is easily excised. In extremely rare cases these lesions are said to take on an aggressive nature and enlarge rapidly, when early surgery is required. I have never seen such a case. Treatment, therefore, is by reassurance, repeated photographic recording for later comparison, and adequate follow-up.

FIG. 1—Large strawberry naevus.

Strawberry naevi are occasionally seen on the lip margin, when feeding difficulties may occur. In these circumstances excision may be justified, but injections of a sclerosant may also produce a satisfying and adequate degree of shrinkage, while natural resolution is awaited.

PORT WINE STAIN—NAEVUS FLAMMEUS

This purely capillary angioma is present at birth. It does not spread, but if on an exposed part can be extremely disfiguring. It can be concealed by appropriate cosmetics (such as Covermark and Keramask) or more permanently by over-tattooing with a light pigment, if the capillaries are placed sufficiently deeply in the dermis to allow room for a more superficial layer of pigment.

In many cases it is possible to excise the affected areas, and close the defect by advancement of local normal skin. If the area affected is large, the technique of serial excision is used. In such a procedure as much of the lesion is excised as can be closed by local skin advancement. After a delay of 18 to 24 months the skin in the area has stretched sufficiently to allow a further part of the naevus to be excised. This procedure is repeated as many times as necessary until the whole has been excised. Free or pedicled skin grafts may be used to replace an excised naevus but in most patients they leave scarring as obvious as the original disfigurement.

CAVERNOUS ANGIOMATA

These venous malformations may be present at birth, or may appear subsequently, as a bluish swelling with comparatively normal skin overlying it. They are compressible, and become more tense when the affected part is dependent. Those referred to the plastic surgeon are usually on the head and neck. Care must be taken to determine the full extent of the lesion by clinical examination, and in some cases by angiography, as extension into the cranial fossa or facial skeleton is not uncommon. In the latter case tooth extraction may be extremely hazardous.

A small proportion of these lesions have an arterial element, which can be detected by palpation and auscultation. Such lesions tend to spread and enlarge. Tying off the appropriate feeding vessels is usually disappointing as further "feeders" develop. Repeated severe haemorrhage from the nose, or other parts, may force the surgeon into radical excision of all involved parts, be they soft tissue or bony. Fortunately such cases are rare.

CYSTIC HYGROMA

This tumour of abnormal lymphatics is rare. It usually presents as a swelling on one side of the neck, perhaps affecting the whole area between the clavicle and the mandible. Infection of the tumour is common and may give rise to respiratory embarrassment. Early excision (in a quiescent phase) is indicated.

LYMPHOEDEMA

A gross reduction in the number of lymphatics draining a limb will cause swelling of the part—usually the lower leg. For some not understood reason this swelling does not usually appear until the teen-ages. Such a swollen limb is sooner or later subject to recurrent attacks of cellulitis and at this point surgery is mandatory. The missing lymphatics cannot be replaced, but excision of the boggy, dripping subcutaneous tissue is helpful and this may be combined with an attempt to link the dermal lymphatics to hopefully unaffected deep lymphatic channels.

PIGMENTED NAEVI

Malignant melanoma is virtually unknown before puberty. Moles that grow hairs medically "never" become malignant. Minor disfiguring naevi on the face may be excised whenever the patient (rather than the parents) request it. Rarely large pigmented naevi covering a fair acreage of the body are seen (fig. 2). While leaving extensive scarring, free skin grafts may well look better than these, and in these extensive lesions the principle of serial excision may also be employed.

FIG. 2—Large hairy pigmented naevus.

Head and Neck

BAT EARS

Protruding ears are often blamed by parents as the cause of their child's lack of success and popularity at school. Undeniably this is frequently true, but equally in many cases a direct question to the patient whether he wants his ears tucked back is met with the answer, "No." Surely it cannot be right to operate on a child merely to please his parents? Better to wait until the child himself requests the operation.

A genuine bat ear has a congenital abnormality, in that the antihelical fold is not properly formed. The fact that the child "always slept with the ear curled forwards" cannot be blamed. At operation the fold is recreated either by force with appropriately placed sutures or by cutting or scoring the cartilage. The incisions, placed behind the ear, do not leave noticeable scars.

OTHER EAR DEFORMITIES

Accessory auricles are comparatively common and often seen, untreated, in adults. Excision in the pre-school period is simple. More severe ear deformities are frequently seen. Where the condition is bilateral, with absent meati, some form of meatal reconstruction is indicated if the middle ear ossicles can be shown radiologically, but in unilateral cases most surgeons consider such an operation unnecessary. Reconstruction of the pinna is difficult. Repeated trauma results in ulceration and exposure of silicone rubber prostheses, so that this material is

largely excluded from use in children. Suitably carved, or otherwise shaped, fragments of costal cartilage may be used, as a framework in the new pinna. At the first operation these are implanted beneath the mastoid skin and later raised from the scalp with a free graft lining the postauricular sulcus.

SINUSES, PITS, FISTULAE, AND CYSTS

Preauricular Sinus

Most commonly seen is the preauricular sinus or "ear pit." This normally presents as a complaint of recurrent abscess formation around the pinna, nearly always at the junction of the upper part of the helix with the temporal skin. Close examination will reveal a small pit, similar to but larger than the punctum of the sebaceous cyst, exactly at this point. The sinus leading from this is variable in length and may branch. It may also communicate with the external auditory meatus. Total excision of the sinus clears the condition, but even after staining the sinus by injected dye, it is difficult for the surgeon to be sure it has all been removed, and recurrence is not uncommon.

Midline Nasal Sinus

This condition also presents with recurrent infection. The opening of the sinus usually lies on the bridge of the nose, in the midline, and a variable distance below the lower tip of the nasal bone. This sinus also may not be easy to excise as it often extends deeply for several centimetres, and may end in a midline dermoid cyst.

Branchial Fistula

A discharging orifice, deeply tethered, on the anterior border of the sternomastoid muscle is diagnostic of branchial fistula. In the quiescent phase only clear mucus is discharged, but during the recurrent episodes of infection to which these patients are liable this changes to pus. Excision is undertaken before school age if infection is troublesome.

Dermoid Cysts

A firm, well-defined, non-tender swelling at one of the described embryological "lines of fusion" suggests the presence of a dermoid cyst. While they may be seen in many sites, by far the commonest is the external angular dermoid presenting at the lateral end of the eyebrow (fig. 3). The calvarium is often

FIG. 3—External angular dermoid.

indented and thinned below the cyst, but rarely deficient. Excision leaves a scar usually placed on the margin of the eyebrow rather than within it, where a bald patch might result.

Many other congenital skeletal and soft tissue facial defects exist. These are all too rare for inclusion here but much can be done to reduce deformity, including if necessary the mobilization of the various elements of the facial skeleton with appropriate repositioning of the bony parts. Nevertheless, one rare deformity does require mention. Coloboma of the eyelid usually results in exposure of the cornea with subsequent drying, ulceration, and perforation, with loss of the eye. Closure of the defect is a matter of urgency.

Limbs

RING CONSTRICTION

The aetiology of congenital ring constrictions (figs. 4 & 5) has been a subject of controversy for some years, but most people think that they are caused by amniotic bands. They may range in severity from minor grooving around a limb or a digit to total intrauterine amputation. If more than mild, the constriction can obstruct the lymphatic return, with corresponding distal oedema. This may often be corrected by suitable surgery to relieve the constriction. When more severe, distal fusion of digits can occur. While no surgeon can produce a normal hand in such patients, much can be done to help, and—as in all congenital hand deformities—the developing child has far more use of his mutilated hand than would be obtained by an adult with the same deformity caused by trauma.

FIG. 4—Mild ring constriction

FIG. 5—Severe ring constriction

SYNDACTYLY

Syndactyly is probably the commonest congenital hand deformity seen in plastic surgery clinics. The most frequent form is fusion between the long and ring fingers. There is usually a positive family history and often the toes are affected as well. Terminal bony fusion may cause lateral deviation of the fingers, owing to differential growth, and this is an indication for early operation. Otherwise the fingers are usually divided at about 4

years of age so that the child has a normal hand to take to school. Invariably there is insufficient skin to cover both digits and free skin grafts are required.

CAMPTODACTYLY

From time to time teen-agers (usually girls) are seen with flexion contractures of the small fingers. This condition also is usually inherited and resists all attempts at surgical correction. There is some evidence that splintage in the early years is effective.

Other hand deformities are rare. Rudimentary accessory digits may be removed at everyone's convenience. Reduplicated digits require the excision of one, the surgeon taking care to leave the better innervated and motivated portion. Absence or under-development of the thumb is often associated with the absence of part of the radius and radial deviation of the wrist. Appropriate splintage may control the wrist deformity, but more commonly internal fixation is required. Transposition of the index finger to the position of the thumb vastly increases the utility of the hand.

Genitalia

HYPOSPADIAS

Two deformities require correction in hypospadias. The ventral bowing of the penis (chordee) must be overcome by appropriate excision of the fibrous tissue on the ventral surface of the penis, and a distal urethra must be created so as to bring the urinary meatus close to the tip of the penis.

There are almost as many operations for dealing with hypospadias as there are surgeons treating the condition. As with any surgical procedure, this indicates that none of them is perfectly satisfactory. The chordee and the urethral deficiency may be dealt with in one stage, or the two deficiencies may be corrected at two different operations. At the time of the urethral reconstruction it is usual to divert the urinary stream, normally by a perineal urethrostomy. This is normally kept in place for seven to 14 days. Fistula formation after the various operations is comparatively common with all techniques and will require further surgery. Spraying of the urinary stream may be troublesome, but infertility is probably not a problem.

Very severe degrees of hypospadias may be associated with

undescended testes and the child may be mistaken for a girl. Chromosomal studies will elucidate matters, but it may well be that if the child has been brought up for some years as a girl, it may be unwise on psychological grounds to alter the assumed sex, and surgery to confirm female sexuality may be indicated. Such cases are rare, but present enormous psychological as well as surgical problems.

CONGENITAL ABSENCE OF VAGINA

This condition may be associated with uterine hypoplasia and usually does not present until puberty. Various techniques exist for vaginal reconstruction, but the simplest of these makes use of a free skin graft to line the newly created pocket. Vaginal delivery after such a procedure has been reported.

EPISPADIAS AND ECTOPIA VESICAE

Ectopia vesicae is normally dealt with by the urologist, but the plastic surgeon may be asked to provide appropriate skin cover after excision of the bladder remnant. Epispadias is extremely rare and involves similar problems of urethral reconstruction as hypospadias.

Trunk

MENINGOCELE AND MYELOMENINGOCELE

In most centres meningocele and myelomeningocele are in the province of the neurosurgeon, but plastic surgeons may be called upon to procure suitable skin cover.

MAMMARY AGENESIS

This condition is usually unilateral and is often associated with maldevelopment of the ipsilateral pectoralis major muscle. Suitable mammary prostheses can produce a considerable improvement, but perfect symmetry is not possible.

I would like to thank the staff of the Photographic Department of the Queen Victoria Hospital, East Grinstead, for the photographs reproduced here.

Conversations on the Social Services

Seaside Problems

FROM A SPECIAL CORRESPONDENT

Dr. Q works in a three-man practice in a south coast town.

"My main criticism about our local social workers is that they don't know their jobs properly—and they're taking a long time to learn. They're lackadaisical about seeing people and have no sense of urgency. They seem unaware that anyone else is able to assess social situations and unaware that doctors usually have a vast knowledge of the family background. Hence if a doctor asks 'move this child' he is probably far more aware of the situation than any social worker

and he usually knows all about deprived children as well. Eighteen months ago I was asked to an inaugural sherry party to meet the new social services set-up. There seemed to be a lot of bewildered inexperienced young people around so I determined that the practice should go easy on its demands, so that they could work themselves in. But things haven't got any easier—in fact the service is definitely worse than before Seebohm.