THE TREATMENT OF SOME SKELETAL DEFORMITIES

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In infants and children there are some rather rare skeletal deformities which are not usually treated by pediatric surgeons and therefore often not treated early enough. As pediatric surgeons have the best opportunities to undertake the care of crippled infants, all diseases and malformations requiring surgical treatment in early infancy should be included within the scope of pediatric surgery.

The very old Chinese custom of binding female feet, and the ritual habits of certain native tribes inducing artificial towerheads, are examples of the fact that treatment in early infancy is capable of moulding the shape of bones during growth. This fact can be made use of in the treatment of certain congenital skeletal deformities. The causes of wrong growth or hindrance of normal growth should also be eliminated as early as possible.

In this lecture I will deal with the treatment of three groups of skeletal deformities: craniosynostosis, thoracic deformities and congenital bone defects of the limbs. Table 1 shows the material treated at our hospital.

Craniosynostosis

The flat bones of the skull are joined by sutures capable of growing like the epiphyseal cartilages in the long bones (Laitinen, 1956). Precocious bony union of the sutures is the cause of craniosynostosis. It results in various deformities of the skull observable soon after birth or detected later.
on account of symptoms caused by increased intracranial pressure.

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The short and broad, high brachycephalic 'tower head' (Fig. 2) is caused by coronal synostosis. As the result of the premature synostosis a disproportion develops between the healthy, rapidly-growing brain and the unyielding constricted skull. This will result in elevation of the intracranial pressure, commonest in combination with the brachycephalic type of craniosynostosis. Persistent pressure may result in retardation of mental and even physical development. The destruction of the optic nerve, leading to atrophy and blindness, is one of the severest complications.

As the growth of both the brain and the skull is maximal in the first year of life and almost complete at the age of four (Fig. 3) treatment at an early age gives the best cosmetic results and affords the best guarantee against any possible brain damage, too. In cases which have been diagnosed later on account of the symptoms caused by increased intracranial pressure the results are also satisfactory (Fig. 4).
Trigonocephaly due to antenatal synostosis of the frontal suture is usually not accompanied by brain symptoms. As the deformity tends to diminish during growth, there is no indication for operative treatment.

Hereditary craniofacial synostosis, the Crouzon syndrome (Fig. 5), usually leads to pressure symptoms sooner or later. Early treatment is indicated.

In Apert's syndrome there is syndactylism (Fig. 6) combined with coronal craniosynostosis (Fig. 7). Opening of the coronal sutures and plastic operations for syndactylism have given satisfactory results in our two patients.

We have treated 82 cases of craniosynostosis by the method recommended by Ingraham and Matson (1954). The results have been satisfactory except for one death due to postoperative shock. One patient, a boy of 2 1/2, had been totally blind for two weeks. The fused coronal and sagittal sutures were opened. After the operation the boy's sight little-by-little returned to normal. The post-operative improvement itself is very good in early cases, but later less good. Fig. 8 shows a child with typical scaphocephaly at the age of 1 month and Fig. 9 the same case after one and a half years. Thus, during the period of maximal growth, i.e. before the age of 4, we consider surgery indicated in every case of craniosynostosis except trigonocephaly. At a later age an operation is indicated only if there are pressure symptoms, because cosmetically it has little effect on the deformity and because attempts to ameliorate the retarded mental status by operation at school age have given disappointing results.

Thoracic Deformities

Deformities of the thoracic wall are rather common in infants and children. Small deformities are unimportant and the indications for...

Fig. 5.—Crouzon syndrome (hereditary craniofacial synostosis).

Figs. 6 and 7.—Apert's syndrome (syndactylism with coronal craniosynostosis).
therapy are only cosmetic or psychological. Deformities involving a great part of the chest wall and sometimes also anomalies of separate bones may to a greater or lesser extent disturb the function of the chest organs. Our experience clearly shows that severe symptoms are rare. Therefore we think that operative correction is indicated only if the treatment is relatively simple and free of risk.

Congenital high scapula (Sprengel's deformity), is often combined with vertebral and costal anomalies. A limited abduction of the extremity is common and the deformity is cosmetically distressing. Operation by the method of Schrock (1926) is easy and gives satisfactory results (Fig. 10).

In antero-lateral defect of the upper ribs the pectoralis muscles are also missing or rudimentary (Fig. 11). The substitution of bone transplants for the missing ribs is recommended, but seldom necessary, because the cosmetic effect is usually due to missing muscles only.

In sternal fissure the upper thoracic aperture is wide, allowing the heart to bulge up to the neck when the child is crying (Fig. 12). Because of cardio-respiratory distress urgent treatment is needed. By freeing the sternal attachments of both sternocleidomastoid muscles and suturing them together in the midline (Fig. 13) displacement of the heart is effectively prevented.

A protruding sternum, or pigeon breast, does not necessitate operation. Marked deformity has sometimes been considered an indication if the operation is free of risk. In cases where the chest wall is depressed on both sides of the protruding sternum cardio-respiratory symptoms tend to be
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Fig. 10.—Result of Schrock’s operation for congenital high scapula.

Fig. 11.—Antero-lateral defect of the upper ribs.

Fig. 12.—Sternal fissure.

Fig. 13.—Correction of sternal fissure.

more marked (Fig. 14). In such cases we have made a subperichondral resection of the depressed costal cartilages. The perichondria are then elevated and fixed to steel struts, which perforate the sternum medially and are fixed laterally to the ribs at the normal level (Fig. 15).

The indications for the repair of funnel chest, the commonest thoracic deformity, are nearly always relative. The operation must therefore be simple and not attended by too much risk. Our operative method earlier consisted of subperichondral resection of the deformed cartilages, retrosternal dissection of the diaphragmatic attachments and maintenance of the elevated position of the sternum with a steel strut passed through the sternum from side to side (Figs. 16 and 17). As we were practically never able to find a real retrosternal ligament and as there is always a certain risk of pleural damage, we further modified our method by abandoning retrosternal dissection. Our last 70 cases have been treated by this simplified method without complications. At follow up the result has been considered good in 80% and satisfactory in 20% of cases.

Congenital Bone Defects of the Limbs

The congenital deformities of the limbs which are caused by primary defects of the bones are
supposed to be due to 'errors of segmentation' as a result of accidents during early embryonic development. Until now they have been rather rare but in recent years their incidence has suddenly risen in some countries. Thalidomide-containing drugs ingested by the mothers during the early stages of gestation are supposed to be the cause of this increase.

The skeletal deformities due to bone defects mainly follow longitudinal divisions of the fetal limb. It is often the case that corresponding parts of the intermediate and terminal segments are lacking.

The therapeutic problems in such cases are difficult. They will be discussed in the following in the light of our clinical experiences and of animal experimentation.

In congenital defect of the fibula (Fig. 18) the
Fig. 18.—Congenital defect of the fibula.

Fig. 19.—Correction of defect of the fibula.

Fig. 20.—Correction of defect of the fibula.
lateral part of the foot is lacking. The ankle is dislocated to the extreme valgus-equinus position. The tibia is both short and angulated. Our four patients have been treated with early tenotomies and osteotomies with satisfactory results (Figs. 19 and 20). During the first post-operative years there has not been any progressive tendency to shortening of the affected legs.

A congenital defect of the tibia may be total or partial. One of our three patients had a unilateral total, one a unilateral partial and one a bilateral partial defect. The varus position was corrected and the ankle stabilized by an operation in which the distal end of the fibula was attached to a drill hole made in the tarsal cartilages (Fig. 21).

Subsequent attempts to join the upper end of the fibula to the epiphysis of the femur in the case of total absence of the tibia resulted in only partial success (Figs. 22 and 23). In partial defect, osteosynthesis between the rudiment of the tibia...
FIG. 24.—Partial bilateral defect of the tibia.

FIGS. 25 and 26.—The final situation in the same case as in Fig. 24.
and the osteotomized fibula succeeded well (Figs. 24 and 25). The girl shown in the figures has since been capable of walking well (Fig. 26). She can also dress herself in spite of the simultaneous hand deformities, due to bilateral radius defect.

Patients with a congenital defect of the femur may be very well helped with a prosthesis only (Grob, 1957). In one of our three cases, with an almost total defect (Fig. 27), we tried to replace the lacking bone with a fibular autograft. The graft survived but showed no evidence of continuous longitudinal growth. In spite of marked shortening of the affected limb, however, the boy is walking and running well with his prosthesis (Fig. 28).

Aplasia and hypoplasia of the radius are the commonest skeletal defects of the limbs. Our material consists of 30 cases. Attempts to replace the missing bone with growing epiphyseal autografts have met with only moderate success (Starr, 1945; Riordan, 1955; Heikel, 1959). Our first method of operation, using epiphyseal transplants of the metacarpal bones (Fig. 29), did not give enough stability to the wrist (Fig. 30). Later, we adopted a new method, which consists of transposition of the ulnar head to a more radial position

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**Fig. 27.**—Congenital defect of the femur

**Fig. 28.**—Result of correction of defect of femur with autograft.
Fig. 29.—Aplasia and hypoplasia of the radius. Operation using epiphyseal transplants of metacarpal bone.

Fig. 31.—Aplasia and hypoplasia of the radius. Transposition of ulnar head.

Fig. 32.—Result of operation in Fig. 31.
against the carpal cartilages (Fig. 31). It has given more satisfactory results with regard to both the cosmetic appearance and the function (Fig. 32).

Our three cases of defects of the ulna were all bilateral and combined with cubital synostosis (Fig. 33). The corresponding fingers were missing. No treatment has been considered necessary.

Subtotal defect of the humerus with corresponding peripheral defects is a type of phocomelia. One of our three cases had a unilateral, the others bilateral defects. One of these also had a simultaneous bilateral radius defect. The rudimentary upper extremity floats unstably in the axillary region. In order to save as much as possible of the upper extremity function we have tried to produce a more stable union between the rudimentary limb and the trunk. We freed the sternal end and the middle part of the clavicle and turned it laterally to the shaft of the extremity. An osteotomy behind the acromion was necessary to achieve this in the oldest child. The clavicle was fixed to the rudiments of the humerus and to the bone of the forearm with wire cerclage (Fig. 34). The capacity of the patients to use their hands has clearly improved post-operatively and the children could now get their hands together in the midline (Fig. 35).

A patient with pure segmental malformation of the forearms and legs of the type described only by Nivergelt, has been seen at our hospital (Figs. 36 and 37). To correct the extreme valgus position, a wedge-shaped resection was performed on both the tibia and the fibula. The epiphyseal lines were turned to a more normal position (Fig. 38). The X-ray four years later shows that there still exists a valgus position of 40 degrees, and therefore a further operation of the same type is planned.

The Growth of Epiphyseal and Sutural Grafts in Animal Experiments

The question of whether it is possible to increase the growth of a hypoplastic bone or to induce significant growth in a transplanted bone is unsolved. In the cases presented above we have obtained some positive results in clinical material. These attempts are based partly on previous publications on this subject, partly on experimental studies on animals. Part of this experi-

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**Fig. 33.-** Defect of ulna combined with cubital synostosis.

**Fig. 34.—** Subtotal defect of the humerus before and after operation.
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Fig. 35.—Result of operation in Fig. 34.

Fig. 36.—Pure segmental malformation of the forearms and legs.

Fig. 37.—Pure segmental malformation of the forearms and legs.

Fig. 38.—Operation on case in Figs. 36 and 37.
mental work, comprising over 100 experiments on growing dogs, rabbits, guinea pigs and rats, was performed by my coworker Dr. Ryöppy.* In the following some examples of our experiments are presented.

The proximal fibular epiphyses of a young dog with a piece of adjoining metaphysis are transplanted from one side to another, thus changing the site only (Fig. 39). In the medullary cavity there is a needle marking the distal point of the graft. In three months the longitudinal growth of the graft is 30 mm. on the right and 17 mm. on the left. The original length of the graft being 8 mm., the growth has been, respectively, about four times and twice the original length. The form of the right fibula is quite normal and on both sides the appositional growth has been normal.

* This part of the experimental material, from which the pictures shown here are also chosen, will be published later by Dr. Ryöppy.

Amputation of the humerus has been performed on a newborn rat (Fig. 40). The distal end of the antebrachium, comprising the epiphyses of the radius and the ulna, has been fixed to the stump of the humerus with a needle. The growth of the graft is evident. In the histological picture the structures of the graft and the host area are seen. There is formation of new bone. The medullary cavities of the graft and the humerus are separated.

Only autogenous transplants have given positive results. Therefore there is no ready source of grafting material in the human subject. In a search for new material for transplantation different animal experiments were performed in which cranial sutures were used as grafts. The membranous bones of the skull grow by proliferation of the sutural connective tissue and secondary apposition of bone at the suture. This process is parallel to the endochondral growth of the long bones.
The type of coronal sutural graft, which has been used in the experiments on the rabbit, is seen in Fig. 41. Such a graft, when reimplanted in its original site, grows in favourable cases in the same proportion as the other parts of the suture. Growth is not very marked, however, because the growth of the skull stops relatively early and because it is very difficult to experiment with newborn animals.

But such a sutural graft, when transplanted to the site of a removed proximal fibular epiphysis, continues its growth in the same proportion as the corresponding fibular epiphysis.
Fig. 42 shows an experiment on a dog. The sutural graft has been fixed in the place of a proximal fibular end removed extraperiosteally. The growth of the graft in three months is two and a half times the original length, corresponding to 25% of the growth of the leg. In the low voltage X-ray of the specimen at the right, the sutural line is visible. At the extreme right of the picture the proximal end of the fibula of the control side is seen.

Fig. 43 shows an experiment on a rabbit. On the right side there is the same type of graft as in the previous case. The removed right fibular proximal epiphysis is transplanted to the corresponding place on the left side. Both the sutural and the epiphyseal graft have grown to three times their original length, corresponding to 50% of the growth of the leg during the same time.

It seems evident that the capacity for growth depends on local factors induced by the host area. Transplantation frequently fails, however. Many questions concerning the factors contributing to the successful results are still unanswered. No definite suggestions for clinical application of these results can yet be given. The treatment of congenital bone defects with grafts capable of growth, however, seems to be within the bounds of possibility.

REFERENCES
