Congenital Anomalies Of The Hand

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THE hand is one of the commonest sites in the body to be afflicted by congenital anomalies which present in a variety of forms. Some of them are so minor as not to produce any functional disturbance, while at the other extreme there may be such severe anomalies causing considerable handicap to the afflicted individual. These children learn to make the best possible use of the deformed hand, and many of them can carry on the normal daily activities with reasonable ease. In the treatment, the functional aspect should always be given priority over the cosmetic aspect, and no tissue that might be functionally useful sacrificed.

CASE MATERIAL

This paper deals with our observations in forty three cases of congenital hand anomalies which presented for treatment at the S. S. Hospital, College of Medical Sciences, Varanasi. The cases have been grouped according to the classification described by Patterson (1959) into six groups as given in table I. Those cases having multiple defects being placed in a particular group according to the most prominent defect present. Twenty two cases had bilateral involvement. Tables II, III, IV and V give the details of the individual cases in various groups.

DISCUSSION

Group I—Webbing of normal fingers :-

Syndactyly is a very common congenital hand anomaly being next in frequency only to polydactyly. Its incidence as reported by Bunnel (1956) varies from one to three per thousand births. Its pathogenesis is not clear but it is believed to be the result of a failure of the webs joining the fingers to disappear during intra-uterine life. Since the thumb is the first to separate, syndactyly of the thumb is least common. Syndactyly shows a strong heriditary tendency, that has been repeatedly traced through several generations.

Syndactyly is frequently bilateral and sometimes the toes may be affected as well. It is slightly more common in males. The middle and ring fingers are the ones most commonly affected. Unless the fussion is producing a deformity of the adjacent finger because of unequal growth, there is no urgency in treatment, which can be done between the age of two to five years. The guiding principles in the treatment of syndactyly are as follows :—

1. Webbing on both sides of a finger should not be released at same operation otherwise there is a risk of ischaemic necrosis of the finger.
2. To attain a normal range of abduc-

tion between the affected fingers, special attention should be paid to the construction of the commissure. Good flap skin is required in this situation which can be provided by : (a) a single dorsal flap, (b) a single palmar flap (Barsky), or (c) two flaps; one dorsal and one palmar.

3. Zig zag incisions are used to separate the fingers to avoid the liability of a straight scar to contract.
4. Since there is a deficiency of skin, additional skin in the form of free grafts is required in most of the cases.

Group II—Webbing of abnormal fingers :

This group includes a variety of anomalies presenting complex appearances. Sometimes the webbing of the fingers is associated with skeletal deformities in the fingers or supernumery fingers. Case one (Fig. 1) in the group had a duplication of the third finger and webbing as well.

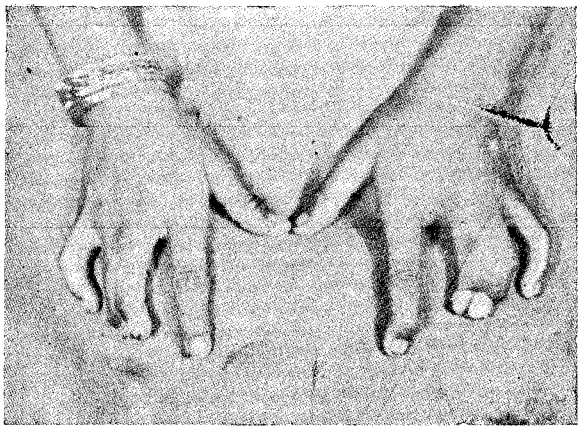

Fig. 1 Showing Fusion of Abnormal Fingers (Bilateral, complete complicated syndactly) involving middle and ring which is duplicated.

The small webbed hand is another example in this group (Fig. 2). It is usually unilateral, the affected limb may be smaller and the sternal head of the pectoralis major may be absent. In females, the breast on the affected side may be smaller than the opposite one.

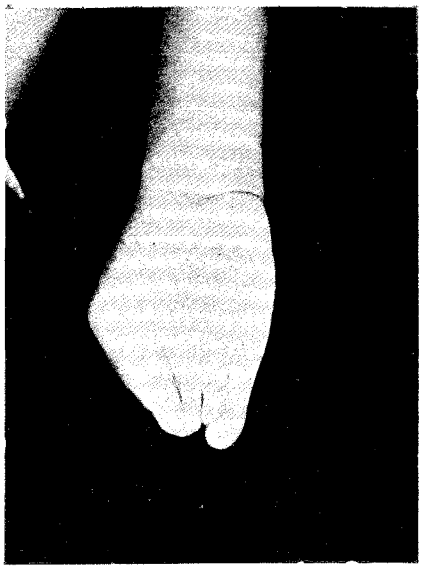

Fig. 2 Showing fusion of abnormal finger (The Small webbed hand.

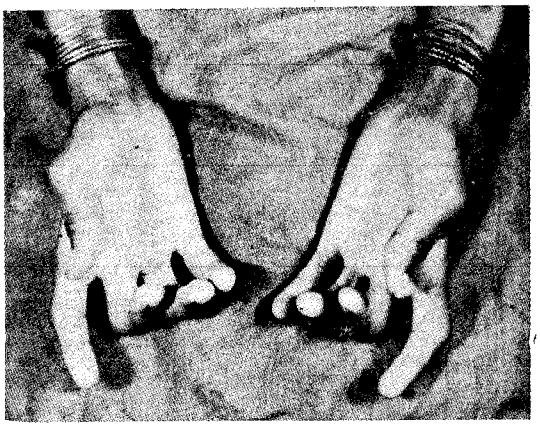

Fig. 3 A congenital flexion contracture involving both hands.

In Apertss syndrome there is a characteristic deformity of the skull associated with a gross fusion of the fingers of both hands and sometimes of the feet as well. There may be a common nail. Function in these

cases is usually poor because of the retarded mental state.

The treatment varies from case to case. The central rays are often underdeveloped and their separation may result in useless flail digits. Sometimes the configuration of the bones and joints is such that simple separation of the webs may not serve any useful function. In the small webbed hand it may be possible to make a cleft and improve the function of the hand. In acrocephalo syndactyly also separation of at least one digit greatly improves function because it provides a useful pinch mechanism.

Group III—Abnormalities of position :— In this group there are mainly two groups of deformities viz. (a) flexion contracture (Fig.3) and (b) lateral deviation.

In mild cases the flexion contracture may often be missed because small infants normally keep fingers in the flexed position. The little finger is the one most commonly affected. The soft tissues on the anterior aspect of the metacarpophalangeal and interphalangeal joints may be contracted. In arthrogryposis multiplex congenita there is a failure of differentiation of muscle as well as contracture of the ligaments and joint capsule. All the four limbs may be affected.

Mild cases of flexion contracture can be treated by corrective manipulation and splinting alone, but most severe cases require operative interference, in the form of a Z plasty followed by splinting.

Lateral deformity when limited to the fingers is known as clinodactyly. This is due to abnormalities of the bone ends forming the joints. More severe lateral deformity at the wrist commonly known as the radial club hand is discussed under group IV.

Group IV—Absence of parts.—The term ectrodactyly denotes the congenital absence of part of a digit or of one or more digits. It may be complete with the absence of all the fingers or one or more metacarpals as well.

On the radial side there are a variety of anomalies. There may be a complete absence of thumbs, only the thumb and the first metacarpal (Fig. 4) or the thumb, first metacarpal and the radius as well (Fig. 5). In some cases the first metacarpal may be missing but a flail thumb may be present and attached a little more distally by a tenuous pedical to the second metacarpal. In mild cases there may be only a hypoplasia of the thenar muscles.

On the ulnar side the ring and little fingers may be absent along with their metacarpals. Ulnar club hand with an absence of the ulna is an extremely rare condition. Fig. 6 shows a case with a congenital absence of the fifth metacarpal.

Sometimes one or more of the central rays may be missing resulting in a cleft hand or a lobster claw hand.

Occasionally there may be an absence of all the digits, or all the parts distal to the carpus may be missing (Fig. 7). The hand may sometimes articulate directly below the elbow or directly at the shoulder with the intervening part of the superior extremity being missing. This condition is known as the phocomelus.

Formerly it was believed that most of these cases were due to intrauterine amputations. But the present day opinion is in

Fig. 4 Congenital absence of thumb and first Metacarpal.

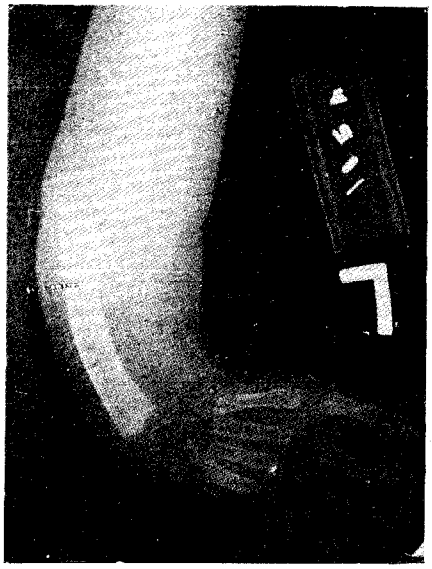

Fig. 5 Skiagram showing congenital absence of radius first metacarpal and thumb.

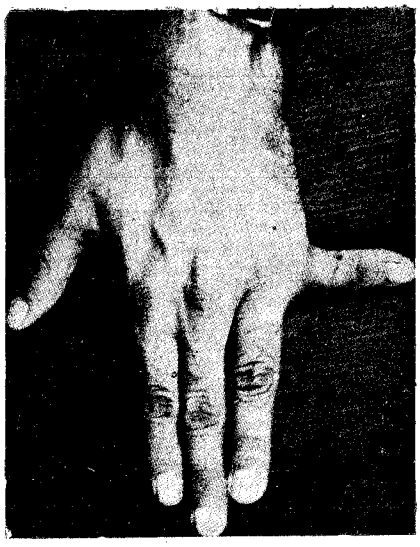

Fig. 6 Congenital absence of fifth metacarpal. The little finger is also attached to the fourth metacarpal.

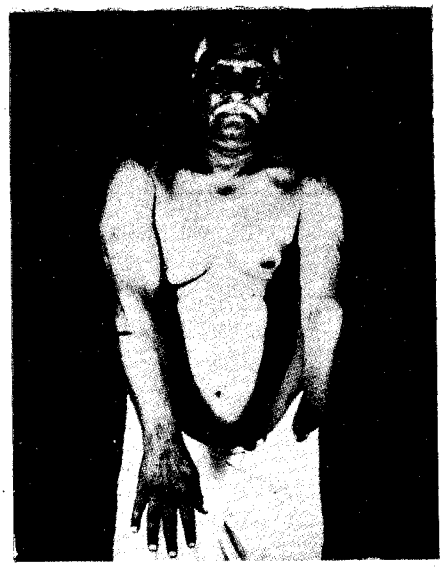

Fig. 7 Congenital absence of the hand distal to the carpus.

favour of the theory of agenesis or failure of development due to some extrinsic or intrinsic factors.

Cases with unilateral absence of the thumb usually develop good function and

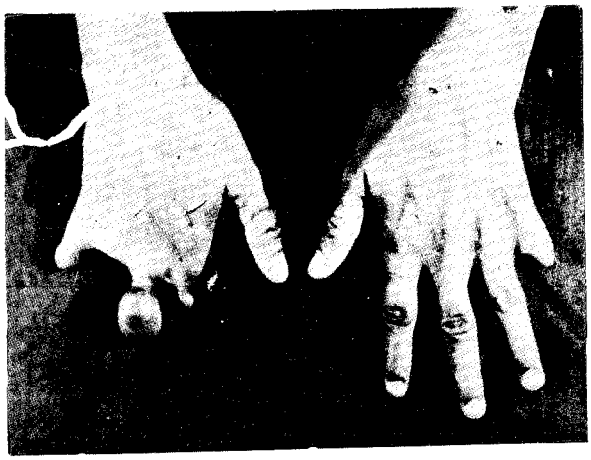

Fig. 8 Showing ring constriction of right middle finger with distal oedema. The congenital amputations of other fingers might be due to ring constrictions during intrauterine life.

the appearance is also cosmetically acceptable. In cases of bilateral absence of thumb, reconstruction by pollicization of the index finger is recommended. In cases of a flail thumb, the pedicle should first be enlarged and later the thumb stabilised by bone grafting.

Radial club hand may be unilateral or bilateral and there may be a partial or complete absence of the radius, along with absence of the thumb and first metacarpal. The hand may assume a position at right angles to the forearm, and in severe cases the radial border of the hand may be along the radial border of the forearm. Soft tissues on the radial side may be contracted and the unla bowed to the radial side.

The treatment of this condition is long and protracted. Entin and Petric (1957) have laid down the principles of treatment for the management of these cases. Corrective manipulation and splinting is started as early as possible with light wire and elastic splints. If this fails to achieve full correction then soft tissue operations to lengthen the contracted tissues on the radial side are done. To maintain the correction achieved, an autogenous bone graft from the proximal fibula is wedged into the ulna to form a buttress for the carpus (Entin 1957, Riordon 1955) at about the age of eight years. Corrective osteotomy of the ulna is done when there is excessive bowing. When full growth of the hand has been achieved bony fixation of the wrist is done by bringing the carpus to the centre of the bony support in the forearm.

For cases of ulnar ectrodactyly no special treatment is required.

Cases with agenesis of the hand or a phocomelus involving both the hands require to be looked after in special homes. Artificial prosthesis can be provided which are quite useful and enable the children to carry out their personal requirements.

Group—V Ring Constriction (Fig. 8)

Annular grooves or ring constrictions are rather uncommon and Patterson (1959) reported 30 cases in a series of six hundred hand anomalies. The constriction may be single or multiple and may affect the fingers, toes, forearm or leg. They vary in depth, the severe ones may reach almost upto the bone and may cause distal lymphoedema etc. Some of the intra-uterine amputations might be due to deep ring constrictions.

Various theories have been put forward to explain the pathogenesis of constriction rings. Amniotic bands were considered to be the cause of the constriction rings, but Streeter (1950) showed that it is more likely that the deformities and the amniotic bands are the result of some developmental failure. He found no evidence that the band is ever the cause of the constriction.

Inglis in 1952 believed that the constriction rings are due to local inferiority of the tissue in various parts of the body resulting from influences transmitted in the germ plasm. Patterson in 1961 put forward the theory that the etiology of constriction rings was analogous to that of cleft lip, that is mesodermal masses under the skin fail to develop resulting in annular grooves.

Shallow grooves which do not interfere with the circulation need no interference. The deeper ones should be excised, preferably half the circumference at a time, and the circular suture line should be broken up by one or more Z plasties.

Group VI—Excess of Tissue : Excess of tissue may be in the form of duplication of digits or a localised gigantism

Duplication of digits is a very common anomaly and is often inherited. In this series two of the cases had a clear family history of this condition on their paternal side and in both cases only the males had been affected. Duplication is most common on the ulnar or the radial side and is rare in between the digits. Duplication of the index finger is extremely uncommon (Hand Forth 1950; Kanavel 1932). Often the anomaly is bilaterally symmetrical and may involve the feet as well. The extra digit may present as a small nubbin of tissue with or without any bone or it may present as a complete fully formed finger with independent tendons, nerves, vessels and bones. A separate metacarpal may also be present. In between these extremes various degrees of duplication may be observed.

Treatment is simple when the duplication is on the ulnar side and usually needs an amputation. In duplications on the radial side, the decision as which thumb to amputate becomes difficult, because neither of them is normal in its size, power or axis. When amputation is done the tendons of the amputated digit may be transferred to the remaining thumb to provide additional motor power to it.

Macrodactyly or gigantism is a rare congenital anomaly and two cases were encountered in this series. The index and the middle finger are most commonly involved. Although all the elements in the hand, including bone tendon and nerves are hypertrophied, the subcutaneous tissues on the palmar aspect seem to be hypertrophied proportionately to a greater degree. This condition is quite distinct from gigantism due to an arteriovenous fistula or neurofibromatosis. Plethysmographic studies in our cases showed a reduced level of circulation in the affected digits. Moore (1944), Mc Carrool (1950) and Tsuge (1967) observed that the digital nerves in these cases are markedly hypertrophied, and Tsuge suggested that the digital nerve anomaly might be the cause of the hypertrophy in the fingers.

The treatment of macrodactyly varies with the degree of hypertrophy, the deform-

ity and the age at which the patient has presented for treatment. Thinning of the digits by soft tissue excision, shortening by amputations, and arresting the growth by epiphyseal ablation are some of the methods of treatment that have been advocated. We are, however, greatly impressed by the operation in which a measuring worm like shortening of the dorsal side is done (Tsuge 1967) Thus the nail is moved back and a considerable shortening is achieved. In children this procedure can be combined with the excision of the distal phalangeal epiphysis.

ETIOLOGY OF CONGENITAL ANOMALIES

The factors of importance in the etiopathogenesis of congenital hand anomalies may be broadly classified as follows :

1. Genetic or intrinsic.
2. Environmental or extrinsic.
3. Complex interplay of both.

Genetic factors are by far the most important. Several long pedigrees can be traced in which the anomaly can be traced through several generations. The genetic factors may be modified by environmental factors. The type of inheritance may be dominant, recessive or otherwise.

Among the extrinsic factors several teratogenic agents such as anoxia, insulin injection, steroids or external irradiation acting during the critical period may produce the congenital hand anomalies in the offspring in experimental animals. In the human being the role of Rubella, irradiation and thalidomide have been clearly associated with congenital hand anomalies. The role of other agents is uncertain. Possibly any of the teratogenic agents acting during the critical period, i.e. the 4th to the 8th week might produce these anomalies. Probably in majority of the cases a complex interplay of genetic and environmental factors may be responsible for such defects in human beings.

Morphogenetically the normal development of the limb depends upon an orderly role of mesodermal condensations in the limb bud and the overlying apical ectodermal ridge, the activity of which is controlled by reciprocal induction mechanism, (Zwilling 1961,Milaire 1962). Anomalous limb patterns can be explained upon atypical distribution or activity of apical ectodermal ridge or the defect of the underlying mesodermal condensation. Deficiency or reduction defects can be generally explained on the basis of loss of activity or degeneration of the mesoderm and/or the ectodermal ridge. Excessive tissue may be explained on the basis of more extensive distribution and/or abnormally prolonged activity of apical ectodermal ridge on the responsive mesoderm.

SUMMARY

A brief review of literature on congenital hand anomalies has been presented along with our observations in 43 cases of such anomalies. The current concepts in the aetiology of congenial hand anomalies have also been described.

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Table I

Showing classification of cases into various groups.

Group I	Fusion of normal fingers	7 cases
Group II	Fusion of abnormal fingers	3 cases
Group III	Abnormalities of position	8 cases
Group IV	Absence of parts	10 cases
Group V	Ring constrictions	1 cases
Group VI	Excess of tissues : Polydactyly	7 cases
	— Gigantism	2 cases
	— Multiple anomalies	5 cases
	Total	43 cases

Table II

Showing the details of cases of fusion of normal fingers.

Sl. No.	Uni-lateral or Bilateral	Whether single, double or tripple	Fingers involved	Partial or complete	Simple or complicated	Involve-ment of feet
1.	Unilat	Single	Middle and ring	Partial	Simple	No
2.	Bilat	Double	Mid ring and little	Complete	Simple	No
3.	Unilat	Single	Mid and ring	Partial	Simple	No
4.	Bilat	Single	Mid and ring	Partial	Simple	No
5.	Bilat	Double	Mid ring and little	Complete	Complicated nails and terminal phalynx fused.	No
6.	Bilat	Single	Mid and ring finger	Complete	,,	No
7.	Bilat	Lt. Single	Mid and ring finger	Complete	,,	No
8.		Rt. Double	Mid ring and little	Complete	,,	No

Table III. *Showing details of cases of group II (Fusion of abnormal fingers).*

Sr. No.	Unilateral or Bilateral	Description	Other associated anomolies
1.	Bilateral	Fusion of 3rd and 4th finger plus one supernumerary 3rd finger present.	—
2.	Unilateral	Small fused hand	—
3.	Bilateral	Complicated fusion of 2nd, 3rd, 4th, and 5th finger.	Apert's syndrome

Table IV. *Showing details of cases belonging to Group IV (Congenital absence of parts).*

Sr. No.	Sub-group	Number	Description	
1.	Congenital absence of parts on radial side	6	(a) Congenital absence of thenar eminence	1
			(b) Congenital absence of thumb and first metacarpal.	2
			(c) Congenital absence of radius	3
2.	Congenital absence on ulnar aspect	1	Absence of fifth metacarpal with attachment of both 4th and 5th fingers to the 4th metacarpal	1
3.	Phocomelia	3	Phocomelia at wrist	1
			Phocomelia below elbow	1
			Phocomelia below shoulder.	1

Table V. *Showing the details of cases belonging to group VI (excess of tissues).*

Sr. No.	Sub-group	Number	Description	
1.	Duplication of thumb	2	Supernumerary thumbs bilateral	2
2.	Ulnar polydactyly	5	Ulnar polydactyly involving both hands and feet	5
3.	Gigantism	2	Index finger	1
			Middle and ring finger	1

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