

CONGENITAL MANUS VARA

By

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The congenital defect of the radius produces a typical deformity of the hand, manus vara. Sometimes the name club hand is also used. Aplasia of the radius with typical deformity of the hand (*Petit*) was described as early as 1733. In 1924 *Kato* stated that he had found 250 cases in the literature and also reported 3 of his own cases. Later a number of publications appeared about this malformation (*Davidsson*, *Goldenberg*, *Heikel*, *Riordan*, *Starr* and others). In *Birch-Jensen's* study from Denmark of congenital deformities of the upper extremities (1949) defects of the radius are stated to have an incidence of 1 in 30,000 births or throughout the whole population 1 in 55,000. A total absence of the radius occurred in 32 of *Birch-Jensen's* 73 cases with defects of the radius. *Heikel* (1959) reviewed 424 previously published cases; total aplasia of the radius was found in 300 of these, including 151 bilateral cases, while a further 31 had total aplasia of the radius on one side and on the other side a partial aplasia or hypoplasia of the radius. Amongst *Heikel's* 47 personal cases there was a considerably lower incidence of total aplasia. Aplasia of the radius can therefore be considered unusual although it is not exactly a rarity.

At the Orthopaedic Clinic in Gothenburg three cases with aplasia of the radius have been treated in recent years.

CASE HISTORY

Case 1: A boy, B.L., born 13/1 1954, appeared for the first time at the Orthopaedic Clinic at Gothenburg in February, 1954, owing to malformation of the arms. He was the youngest of nine children in a family with no known malformation or blood disease. The boy's two arms were obviously short, especially the forearms. His hands were severely bent volarly and radially, and both thumbs turned inwards. No radius could be observed on the X-ray. The mother was instructed to

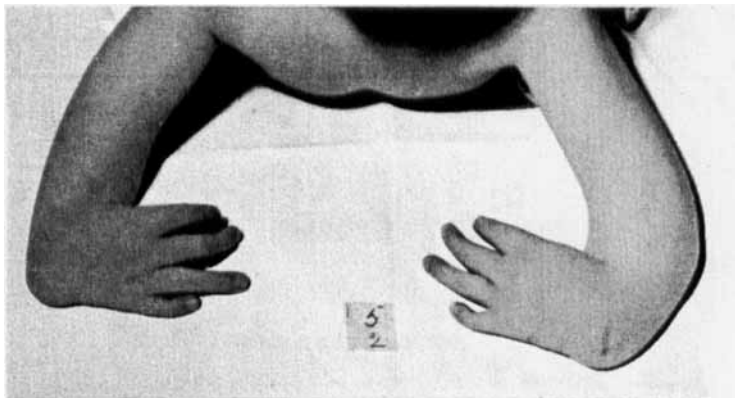


Fig. 1.

Case 1. Boy born 13/1, 1954. Picture in April, 1956 before treatment.

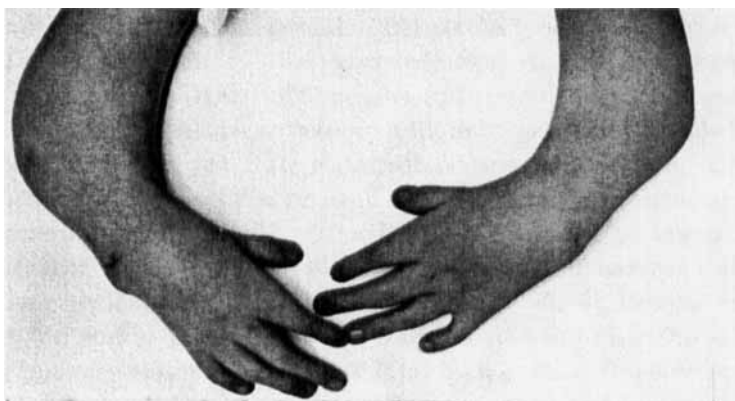


Fig. 2.

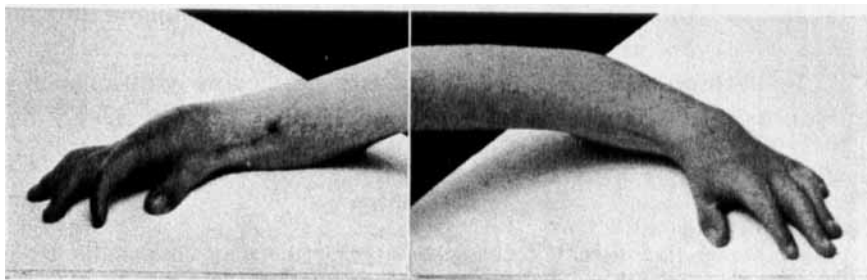


Fig. 3.

Fig. 4.

Figs. 2, 3 and 4. Case 1, after corrections and operations.

Fig. 3. Lateral picture, right hand. *Fig. 4.* Lateral picture, left hand.



Fig. 5.



Fig. 6.

Figs. 5 and 6. X-ray in April, 1956, forearms. Case 1. The radius is absent bilaterally.

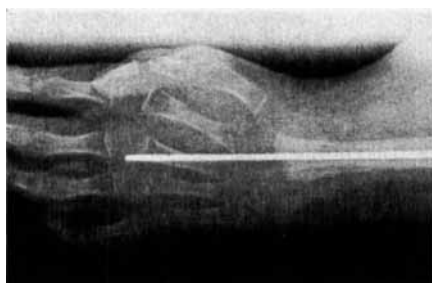


Fig. 7.

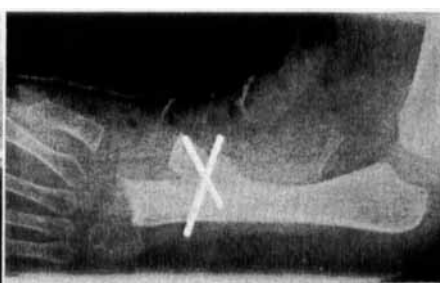


Fig. 8.

Fig. 7. X-ray in January, 1957, left forearm. Case 1. The hand was reduced openly and the carpus fixed to the ulna with a Kirschner wire.

Fig. 8. X-ray in June, 1957, left forearm. Case 1. The transplanted end of fibula is fixed to the ulna with two crossed Kirschner wires.

stretch the boy's hands in the corrective direction and also to try and persuade him to use the hands actively. In April, 1956, at a control examination his ability to grip was relatively good, and he could also feed himself. The deformity of the hands was unchanged. (Fig. 1). Passively the deformity could only be partially corrected. Both passively and actively a moderate limitation of movement was present in the joints of the fingers. Active flexion was found in all fingers but there was no evident extension. Treatment was initiated with correction under narcosis and with plaster in stages so as to prepare for surgical intervention. On examination of the blood, thrombocytopenia was ascertained (74,000 and 24,000 thrombocytes per mm.³ in two cases), but no anaemia was present and bleeding and coagulation times were normal. Owing to the thrombocytopenia the operation was postponed for a time.

When aged 3 (30/1, 1957) the patient underwent open reduction of the ulna to-



Fig. 9.



Fig. 10.

Fig. 9. X-ray in August, 1958, left forearm. Case 1. The fibula transplant has well healed with the ulna which has grown.

Fig. 10. X-ray in April, 1959, left forearm. Case 1. The fibula transplant seems on the picture to have increased in length and an epiphysis nucleus has emerged. The ulna has grown both in width and length.

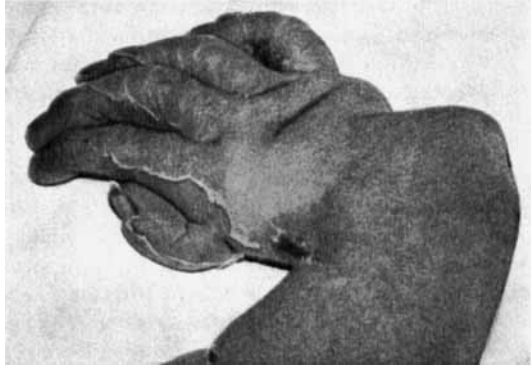
wards the carpus + fixation with Kirschner wire according to Riordan. (Fig. 7). Three months later the second stage of the operation was performed, transplantation of the upper part of the right fibula to the ulna. (Fig. 8). At the operation the epiphysis of the fibula was also taken and the fibula transplant was inserted into the ulna and fixed with two Kirschner wires crosswise through the transplant and the ulna. The post-operative course was uneventful.

On the 9th of May, 1958, the first operation on the right hand was performed. Before the operation he had 54,000 thrombocytes per mm^3 , otherwise the blood tests were normal. Some hours after the operation a quickly-passing rash resembling rubella appeared on the body. The rash was considered to be an allergic symptom. Three days later there was nose bleeding with suddenly considerable blood-vomiting; shock occurred. Blood transfusion and treatment with adrenocortical steroids were given immediately. The condition improved rapidly. The thrombocyte count which on bleeding was 9,000/ mm^3 rose as high as 190,000/ mm^3 but later dropped again to about 50,000/ mm^3 when the steroid dose was reduced.

On 29/8, 1958, the second stage of the operation was performed on the right hand without complications. Then the patient was given prophylactic treatment with adrenocortical steroids. (All operations were performed by Professor E. Severin).

At the last check-up in April, 1959,—at 5 years of age—the boy had good function in both hands with active flexion and extension ability in all the fingers, although there was a certain limitation of movement in the joints of the fingers actively and passively, approximately as before the operation. The cosmetic result was good. (Figs. 2, 3 and 4). X-ray examination of the left hand showed a well-healed transplant, which on the X-ray had increased in length, but whether this is true growth or progressive ossification of the epiphysis cannot be determined. An epiphyseal nucleus had emerged in the transplant. The ulna had grown in length and breadth. The hand is situated in position to the middle of the end of the ulna. (Fig. 10).

Case 2: A boy, B.S., born 15/3, 1958, was shown at the Orthopaedic Clinic as a newborn. There was no known malformation or blood disease in the family, where

*Fig. 11.*

Case 2. Boy born 15/3, 1958. Photograph of right arm in March, 1958.

*Fig. 12.*

Case 3. Woman born 4/10, 1935. Photograph in September, 1959.

he was the second of two children. His right hand was severely bent volarly and radially. The thumb which was rudimentary and connected only by a thin stem, lacked function. (Fig. 11). Active power of movement was found in the remaining fingers. The right forearm was shortened. The X-ray examination of the right forearm showed that the radius and first metacarpal were missing. The base phalanx of the thumb was very small. There were no other skeletal malformations. The blood values were normal apart from a temporary thrombocytopenia. Later examinations repeatedly demonstrated a normal thrombocyte count. The thumb became gangrenous owing to the fact that its thin soft tissue shaft became twisted in one night. The thumb was removed. Then treatment was initiated with corrections and plaster by stages and the same operative interventions were performed as reported above in case 1.

Case 3: A woman, E.J., born 4/10, 1935. According to her own information she was

brought at the age of 2-3 to the Orthopaedic Clinic in Gothenburg for her malformed hands. No measures were undertaken. The record cannot be found, however, and her information is vague. Possibly the patient was instead examined elsewhere. In May, 1959, she was examined at the Orthopaedic Clinic. There was no known malformation in the family. In 1955 she underwent a colostomy for congenital malformation of the rectum.

Arms: Pronounced congenital malformations. Her right upper arm was of normal length but the left upper arm was very short (10 cm.). Her forearms were short (about 14 cm.). Her hands showed a severe varus position and the thumbs were missing. (Fig. 12). There was considerable limitation of movement in the elbow joints and stiffness in the wrist joints. Mobility in the fingers was poor. The hand function was astonishingly good but the strength was poor. She could fasten buttons and grip objects. She fed and took care of herself.—For the time being she is being trained as a photographer at the Anggården craft schools.

The blood tests showed no anaemia or thrombocytopenia.

The X-ray examination of the forearms showed that the radius was completely absent and also some of the carpal bones and the first metacarpal bone.

No treatment of the patient's malformations is planned.

DISCUSSION

In congenital aplasia of the radius the hand has a varus position. The forearm is shortened and sometimes short upper arms are also seen, so that one may even speak of phocomelus, which *Hill* has described. In one of my cases a very short upper arm occurred on one side. In 50 % the malformation is bilateral and if unilateral, the right side is much more commonly involved than the left side (*Riordan* and *Heikel*). Other skeletal malformations are often present. Thus the first metacarpal bone and a number of carpal bones may also be missing as well as the thumb, which sometimes may also be rudimentary and useless. The muscles of the hand and arms may be defective or completely absent. Aplasia of the radius is found in combination with malformations of varying types, often serious defects, so that the prognosis is often poor, but those who survive childhood probably have possibilities of a fairly normal life (*Birch-Jensen*). In a number of cases with aplasia of the radius blood changes occur such as Fanconi's syndrome and thrombocytopenia (*Riordan* and *Gross* etc.). In two of the author's cases thrombocytopenia appeared. It was possible to carry out surgical treatment in case 1 in spite of a pronounced thrombocytopenia. With respect to the combination of aplasia of the radius and thrombocytopenia the reader is referred to an article to be published in *Acta Paediatrica* (*Nilsson* and *Lundholm*).

The etiology of aplasia of the radius is unknown. The malformation

may be inherited dominantly and apparently also recessively, but in the majority of the cases no hereditary character appears (*Birch-Jensen*). In none of the author's three cases was a malformation known to be present in the respective families.

TREATMENT

Several different methods of treatment for congenital manus vara have been described. The malformation must be corrected surgically, because corrections, plaster and splints do not in the long run produce lasting results. However, as a rule conservative treatment precedes the surgical interventions.

Lange suggests conservative treatment up to three years of age, when soft tissue operations with tendon lengthening are performed. If the surgeon is unsuccessful in obtaining a good position of the hand a shortening osteotomy is later performed on the ulna. Various suggestions have been made for maintaining the hand in position. *Lange* considers that the final operation should not be performed until 14 years of age. According to *Bardenheuer* the ulna is divided distally and the carpal bones are placed between the two ends of bone. *Reyerson* placed the rear part of the ulna in the radius position. A tibial transplant is used according to *Albee's* method, which shows good results primarily, but when the bone transplant does not progress with the growth of the ulna, recurrence may occur. In order to avoid this a resection of the epiphysis of the ulna can be performed but this results then in diminished growth of the already short forearm. *Campbell* makes a hole in the carpus and implants the end of the ulna in this. In *Gocht's* procedure the distal end of the ulna is wedged centrally into the carpal bones. The hand may also be fixed to the ulna with marrow nailing according to *Stracker's* proposal.

Riordan describes a plan of treatment in which he, like *Starr*, employs the proximal part of the fibula with its epiphysis, placing them in the position of the radius. *Riordan's* method distinguishes itself from *Starr's* in that in the first stage of the operation the carpal bones are placed in a normal position for the distal end of the ulna of open reduction and are possibly fixed there by Kirschner wire. The operation is performed through an incision from both ulnar and radial aspects. With a curvature of the ulna an osteotomy is performed later if this should prove necessary. In a later stage of the operation the upper part of the fibula is placed to support the carpal bones radially and the in-

tention is that the fibula with its epiphysis shall grow in such a way that no recurrence appears when the ulna grows.

Riordan in his cases did not see any certain growth of the transplanted bone, but increase in the length of the transplant on the X-ray was stated to be the result of ossification of the cartilaginous epiphysis. Transplanted epiphyses have, however, been observed to grow. In operated cases of aplasia of the radius *Heikel* and *Bintcliffe* have observed growth in the transplanted fibula end. *Riordan* considers that the earlier the transplantation of the fibula, the greater are the possibilities of true growth. *Riordan* found no drawback in removing the upper end of the fibula, but one of *Heikel's* operated cases resulted in a valgus position of the foot owing to displacement of the fibula in the proximal direction. In order to avoid this tendency of the fibula to slip upwards *Heikel* recommends an osteotomy on the upper part of the fibula and has the bone end placed into the tibia, so that a bony bridge may then be obtained between the proximal end of the fibula and the tibia. In the first of the author's cases no tendency towards increased valgus loading of the feet was seen.

In *Riordan's* 4 operated cases with total absence of the radius, 2 were followed-up for respectively 3 and 5 years and proved to be successful. His third case had to be re-operated, however, since the end of the fibula had to be replaced in the ulna again. The X-ray examination showed when *Riordan's* article was written that the fibula was now firmly healed in the ulna. The fourth case was unsuccessful because the transplant was accidentally destroyed. Later this case was re-operated according to *Bardenheuer's* method. It is still too early to judge definitely the result of *Heikel's* two operated cases of total radial aplasia with transplantation of the fibula, but the position of the hand seemed to be improved and the mobility to be fairly good.

In the bilateral case described by the author the X-ray picture of the hand operated on first shows an increase in length of the bony shaft section of the transplant and an epiphysis nucleus has also emerged, but it is still too early to decide whether it is true growth or only an ossification of the cartilage. (Fig. 10). Both functionally and cosmetically the result of both hands was successful in this boy. (Figs. 2, 3 and 4).

The treatment of manus vara congenita should be commenced as soon as possible. First stretching is practised, then correcting, plaster, possibly under narcosis. If, at an early age, the hand is given its correct position, better function is achieved, since the muscles can work under

more favourable condition. The operative interventions must not be delayed, therefore, but they must be carried out quickly when the best possible position has been achieved with conservative treatment. Before the operation one must make certain that the patient has no other complaint which might create difficulties for the operation. The possibility of pathological blood changes ought then especially to be kept in mind. As a support for the carpal bones instead of the radius the upper part of the fibula with its epiphysis is recommended.

S U M M A R Y

An account is given of three cases with congenital aplasia of the radius, two children and a woman. Two were bilateral, two were combined with thrombocytopenia. The etiology of this unusual complaint is unknown.

In aplasia of the radius the hand is in the varus position—*manus vara*. Various methods of correcting the malposition are studied. The treatment of a child with bilateral changes obtained good results from corrections, open reduction and finally transplantation of the upper part of the fibula as a support for the carpus on the radial aspect. The treatment of a child with unilateral change has commenced.

R E S U M E

Compte rendu de trois cas d'aplasie congénitale du radius, deux enfants et une femme. Les deux étaient bilatérales et deux étaient combinées avec une thrombocytopénie. L'étiologie de ces cas exceptionnels est inconnue.

Dans l'aplasie du radius, la main se trouve en position varus—*manus vara*. Différentes méthodes de correction de la déviation sont examinées. Un enfant ayant des modifications bilatérales a été traité avec de bons résultats par des corrections, une reposition ouverte et finalement la transplantation de la partie supérieure de la fibule comme support pour le carpe du côté radial. Le traitement d'un enfant ayant une déformation unilatérale a été entrepris.

Z U S A M M E N F A S S U N G

Bericht über drei Fälle mit kongenitaler Radiusaplasie, zwei Kinder und eine Frau. Zwei Fälle waren doppelseitig und zwei waren kom-

biniiert mit Thrombocytopenie. Die Äthiologie dieser ungewöhnlichen Erkrankung ist unbekannt.

Bei der Radiusaplasie steht die Hand in Varusstellung – manus vara. Die verschiedenen Methoden für die Korrektur der Fehlstellung werden durchbesprochen. Ein Kind mit doppelseitigen Veränderungen wurde mit gutem Resultat mittels offener Reposition und folgender Transplantation des proximalen Fibula-anteiles zur Stütze der Handwurzel auf der Radialseite behandelt. Die Behandlung eines Kindes mit einseitiger Veränderungen wurde in Angriff genommen.

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