Acta orthop. Scandinav. *XXXV*, 202–211, 1965.

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# CONGENITAL ANKYLOSIS OF THE ELBOW

## *Late Results in one Operated Case with Bilateral Ankylosis*

*By*

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Congenital ankylosis of the cubital joint is extremely rare. When reporting one case, *Romanus* (1933) had been able to collect only 23 cases formerly described in the literature. *Frostad* (1940) reported five cases of bilateral ankylosis in adults. They all came from two families who lived in the same district (Møre), but without known relationship between the families.

This author (1946) reported on bilateral cubital ankylosis in two baby sisters from the same district as *Frostad*'s patients. The grandfather of the two children had three brothers suffering from the same disorder. The case to be reported here apparently occurred quite isolated, with no family history of similar cases and living far from the district of the other reported cases (Vestfold).

### *Other Malformations.*

In the cases reported earlier by *Frostad* and the author, no malformations other than the ankylotic elbow were seen.

In several of the cases collected from the literature by *Romanus*, however, also other defects had been described in the hands, fingers or fore-arms, such as concrescence of metacarps, absence of metacarps or fingers, proximal or distal ulnar defects and shortening of the fore-arm or the upper arm. In *Romanus*' own case the affected arm was half as long as the sound arm.

In the present case, also cubital ankylosis was one of several congenital defects: Ankylosis of tarsal joints with varus deformity of the feet, symmetrical absence of the first interphalangeal joint in 2.–5.

fingers and ankylosis of the carpal joints. The ankles and wrist joints were normal.

*Anatomical Findings.*

In all of the formerly reported cases, except one, the radius had been found to be the chief bone in the formation of the ankylosis, merging broadly with osseous connection directly into the humerus. In one case, the ulna was ankylotically joined with the humerus, while the radius articulated freely with both. In all the cases reported from Norway (eight cases, including the present case), the radius has formed a broad ankylosis with the ulna and has been the dominating bone of the fore-arm. Of the two cases formerly described by the author, one had no ulna at all. In both cases the malformation was bilateral.

In none of my three operated cases (6 elbows) could epiphysial lines be demonstrated in any of the bones at the elbow. The brachialis muscle was so poorly developed, that its presence at operation could not be decided with certainty. Biceps, the long head of triceps and the muscles of the fore-arm originating at the humerus, were all present. This finding is not surprising, as these muscles also exert their effect on the shoulder and the wrist joint, respectively, and thus maintain some activity despite a stiff elbow.

The presence of these muscles is of the greatest importance to a successful operative treatment of a congenital ankylotic elbow.

*Treatment.*

In schools of surgery and in textbooks it seems to be an established rule that in cases of congenital ankylosis of the elbow, arthroplasty should be postponed until the patient reaches adult age, partly because of the close relationship between the epiphysis and the joint, but especially because of the difficulties connected with the after-treatment in children. The finding of absent epiphysial lines seems to cancel the first problem. It is the aim of this report to demonstrate that the second problem may also be overcome.

The need for treatment is made evident by *Frostad's* description of his five adult patients: "Certainly the ability to work is surprisingly good, even in occupations demanding great exertion, but there are a number of important, daily tasks which these five patients cannot perform. For example, they cannot wash the face or neck, the upper

part of the chest or back. They cannot shave, wash their teeth or comb their hair. They cannot button of their coats or the upper part of the vest. They all have difficulties in eating. They have to bend the head forward in order to meet the spoon or fork."

Faced with these adult patients, *Frostad* took a rather pessimistic view of the possibility of restoring good function by operative treatment: "The possibility of fashioning a new joint by operation has been discussed but not tested. It must be assumed that an attempt to do so would be rather fruitless—chiefly because the muscles that should move the joint are completely atrophied."

When I undertook to operate on the two children in 1946 (an operation which I could not see had been done before in similar cases), it was because I felt that the operation should be performed in early childhood, before inactivation atrophy of the muscles had time to occur. I considered this factor important enough to justify operation despite the considerable technical difficulties met with in these very young patients, as well as the problems encountered in after-treatment when active exercises of the muscles are mandatory.

The results in these two first operated cases were discouraging: After four years one elbow in each patient had again become stiff, the other showed a movement from 90°–130°. The results were identical in both patients.

The result in the present case was more encouraging, and will be described below.

## CASE REPORT

A 4 months old girl with bilateral, congenital ankylosis of the elbow was admitted to the hospital in 1950. She also had other congenital defects of bones and joints, which have been described above. Roentgenogram (Fig. 1) of the left elbow revealed a broad synostosis between the radius and the humerus. The ulna was separated from both humerus and radius by a well defined slit. The olecranon was absent. In the right elbow (Fig. 2) there was a broad synostosis between the humerus and both the radius and the ulna, the latter being very poorly developed. No epiphysial lines were present in any of the bones at the elbow. Despite this fact, she had no marked shortening of the arms. Possibly, a compensatory increase of growth may have occurred at the upper epiphysis of the humerus and the lower epiphysis of the radius.

1) On Feb. 14, 1950, an *arthroplasty of the right elbow* was performed, using the dorsal, longitudinal incision of Langenbeck (this incision has been used in all my cases). A free graft of fat from the abdominal wall was used as interposition material. In accordance with the findings at the roentgenogram, a solid, bony connection between the ulna and the humerus was also found at operation. From the

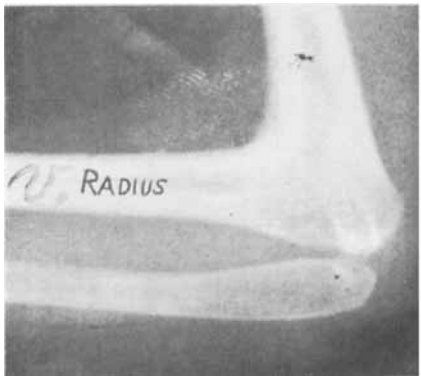


*Fig. 1.*
Congenital, bilateral ankylosis of the elbow. Roentgenogram of the left elbow at age four months. At operation at the age of one year (arthroplasty) was found a cartilaginous connection between the ulna and the humerus. A trochlear notch was fashioned on the ulna and a matching trochlea on the humerus.

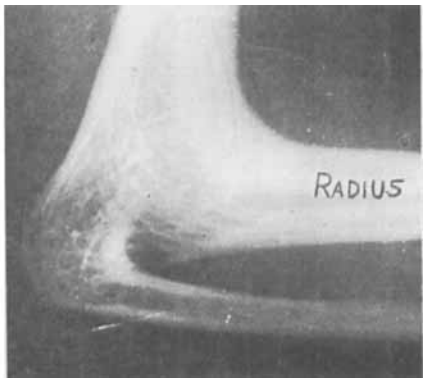


*Fig. 2.*
Roentgenogram of the right, ankylotic elbow at the age of four months, just before the operation, which was performed when the child was 4½ months old. Two re-operations had to be performed to obtain the result shown in Fig. 3.

roentgenogram one may be led to the conclusion that the proximal end of the radium is cylindrical, as in normal cases. As in most of these cases, however, the bone at operation was found to be flattened from one side to another, like a comb, and was sclerotic and very hard to chisel. The wound healed primarily, and the patient could leave the hospital, after the mother had been instructed how to exercise the elbow at home. On doing this, she noted after a while that the child cried during the elbow exercises, but she continued the manipulations as before until one day a marked swelling and stiffening of the joint developed, evidently due to hemorrhage. No fracture could be demonstrated, but the exercises had to be cancelled and ankylosis was re-established in a 110° flexion position.

2) On June 30, 1950 (the child was now one year old) an *arthroplasty of the left elbow* was performed. The synostosis between the radius and the humerus was chiselled through and about 15 mm of the proximal end of the radius resected. The volar prominens at the lower end of the humerus was chiselled off. A cartilaginous connection was found between the ulna and the humerus. A trochlear notch was fashioned on the ulna, and a trochlea to match on the lower end of the humerus. A graft of fascia from the thigh was sutured into the new joint with catgut. After the operation the elbow could be flexed 90° + 20° and be fully extended. The wound healed primarily.

3) On June 28, 1957 (seven years after the first operation, the patient aged 8 years) a *second arthroplasty of the right elbow* was performed. As interposition material this time was used a full skin graft (according to Kallio). After the operation the elbow could be flexed 90° + 40°. The operation had been performed with-

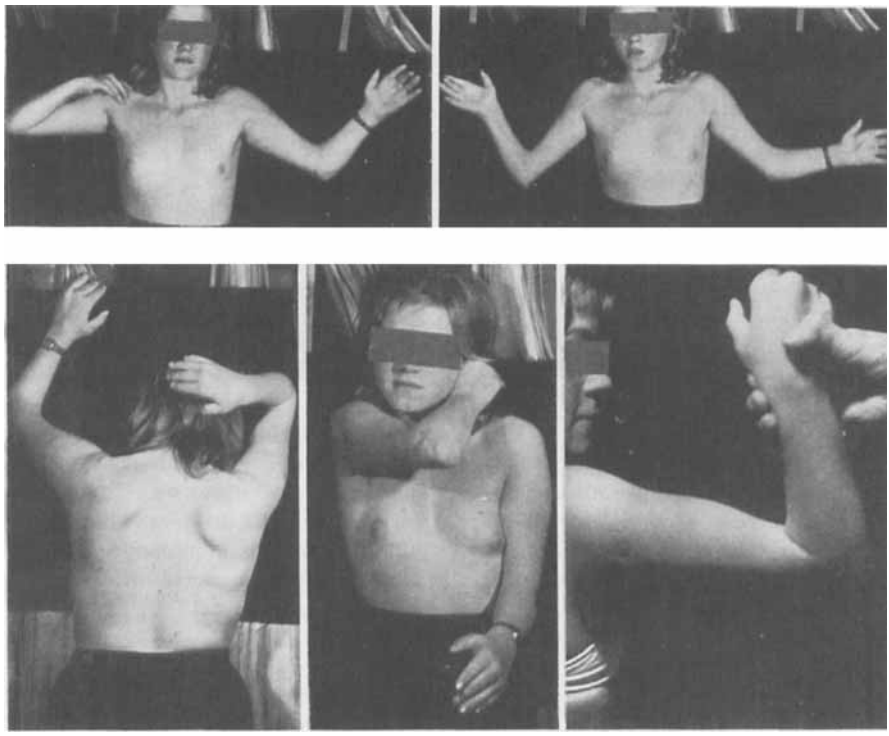

*Fig. 3.*
Congenital, bilateral ankylosis of the elbow, 13 years (left elbow) and 6 years (right elbow) after operation. The right elbow was operated three times because of relapse of the ankylosis. Flexion: L. 85°, R. 90°. Extension: L. 90° + 35°, R. 90°. Note how well the movements of one arm fill the lack of movement in the other: With the right hand she can reach her head and upper part of the body, with the left the lower part of her body. In one picture the bulk of the biceps is shown under flexion against resistance.

out using a tourniquet, and a p.o. roentgenogram revealed a small fragment of bone lying in front of the radius, which had not been discovered at operation. This bone fragment later joined in osseous connection with both the radius and the humerus, and ankylosis was re-established.

4) On October 10, 1957 (four months after the last operation) a *third arthroplasty of the right elbow* was performed. A solid, osseous connection between the radius and the humerus was found at operation. Only a small piece of the full skin graft from the last operation could be recognized, which on microscopy showed "necrotic skin with rejected epithelium". About 12 mm of the proximal end of the radius was resected. As interposition material a flap of fat from the abdominal wall was again uséd, and sutured in place with catgut. Postoperatively the patient, at the age of eight years, could be persuaded into active exercises, and after two


*Fig. 4–A.* Roentgenogram of the right elbow in maximal flexion after the last operation at the age of eight years. The elbow was operated on the first time at the age of 4½ months.

*Fig. 4–B.* The same as in Fig. 4–A, but with the elbow in maximal extension.

months she could actively bend her elbow 90° + 60° and extend to 90°. She could pronate 40° and supinate 15°.

On *follow up examination,* October 1960, 13 years after the operation of the left elbow and 6 years after the last operation of the right elbow was found (Fig. 3):

Right elbow: Flexion 90° + 70° – extension to 90° (Fig. 4 A and B).
Left elbow: Flexion 85° – extension 90° + 35° (Fig. 5 A and B).

The roentgenogram of the left elbow shows that flexion could have been improved by trimming the proximal end of the radius which is pushed against the lower end of the humerus in maximal flexion of the joint. However, the patient was quite content with her elbow and wanted no more operations. The change in the mutual caliber of the two bones of the fore-arm towards normality was remarkable. It may further be seen from the pictures in Fig. 3, how well the movements of one arm fill the lack of movements of the other, thus enabling the patient to meet the basic needs of daily life: The right arm has normal flexion, but cannot be extended past 90°. The left arm has flexion


*Fig. 5–A.* Roentgenogram of the left elbow, 13 years after operation (operated at the age of one year). Flexion, which is limited by the proximal end of the radius pushing against the humerus, could have been improved by another operation. Note change of the mutual size of the fore-arm bones towards normality.

*Fig. 5–B.* The same as in Fig. 5–A, but with the elbow in maximal extension. Extension is limited by the ulna (olecranon) pushing against the dorsal aspect of the humerus, and could have been improved by another operation. The patient, however, is quite content with the arm and wants no more operations.

only to 85°, but may be extended to 125°. She thus is able to reach her mouth, neck, both ears and upper part of her body with her right hand. With the left she can reach the lower part of her back and abdomen with the genitals.

## SUMMARY

The author in 1946 reported on two cases of congenital, bilateral ankylosis of the elbow, which he had operated in 1943. The results were rather discouraging: Of the four operated elbows, two (one in each patient) had relapse of ankylosis, two had a maximal movement of about 30°.

A report is given on one patient with congenital, bilateral ankylosis of the elbow, who was operated for the first time in 1950, at the age of 4½ months, when arthroplasty was performed in the right elbow. This elbow had to be re-operated twice, the last two operations being performed at the age of eight years. The left elbow was operated at the age of one year, and no re-operations were necessary.

Follow up examination 13 years (left elbow) and 6 years (right elbow) after the operation showed rather encouraging results: The patient could make very good use of her arms and was able to meet nearly all the basic needs of daily life. The results are demonstrated by photographs and roentgenograms.

The optimal time for operation is discussed: Whether to operate in early childhood in spite of the difficulties encountered at this age at operation and in after-treatment, in order to avoid inactivation atrophy of the muscles which would be expected at an older age in unoperated cases. Or to operate when the child has become older, when better anatomical conditions at operation and better cooperation from the patient during after-treatment may be expected.

The author has operated on all his cases (three patients, six elbows) in early childhood, because he felt that avoidance of inactivation atrophy was of paramount importance. However, after observing good results after re-operation of a re-ankylosed elbow at the age of eight years, the author now feels that operation at this age may be at least as good as in early childhood.

The occurrence of inactivation atrophy of the muscles around the elbow may have been overestimated, especially of the muscles that in addition to the elbow exert their action upon the neighbouring joints (wrist and shoulder).

The author advocates that operative treatment be considered in all cases of bilateral, congenital ankylosis of the elbow, no matter what the age of the patient.

RESUME

L'opération pour l'ankylose congénitale du coude n'avait pas été mentionnée dans la littérature lorsque l'auteur communiqua en 1946 deux cas bilatéraux opérés en 1943. L'auteur n'a pas non plus trouvé plus tard de communications sur des cas de cette maladie qui auraient été opérés.

Les quatre articulations du coude opérées en 1943 ont en ce qui concerne les articulations du coude gauche chez les deux malades don-

né un résultat négatif avec raidissement. L'articulation du coude droit a chez les deux malades conservé une mobilité limitée de 30 degrés. Elles ont toutes été opérées à l'âge de nourrisson.

Le troisième cas que l'auteur a opéré en 1950 et qui est rapporté ici a donné un bon et durable résultat, très utile pour le malade. Chez ce malade, l'articulation du coude droit a été opérée alors que la fillette était âgée de 4 mois et demi, le coude gauche alors qu'elle avait 12 mois. Il a fallu réopérer le coude droit 3 fois, la dernière fois lorsque la fillette était âgée de 8 ans. Le résultat après 6 et 13 ans respectivement est décrit et il en est fait la démonstration par des illustrations et des radiographies.

L'auteur discute la question de savoir s'il convient d'opérer à l'âge de nourrisson, alors que l'on peut supposer que l'atrophie d'inactivité des muscles n'est pas encore très prononcée ou s'il faut attendre que l'enfant soit un peu plus âgé, alors que l'opération est techniquement plus facile à exécuter et que les conditions du traitement post-opératoire sont plus favorables.

En ce qui concerne l'atrophie d'inactivité, l'auteur pense, en se basant sur les expériences du dernier cas, qu'elle n'augmente apparemment pas très rapidement dans les muscles des trois éléments de l'articulation si importante pour le pouvoir de flexion et de tension du coude. Le résultat chez cet enfant de 8 ans – il est vrai réopéré pour la troisième fois dans l'articulation du coude droit – semble montrer qu'il est possible qu'une opération à cet âge soit plus favorable.

Dans tous les cas, il ne convient pas de rejeter le traitement chirurgical comme n'ayant aucun espoir de réussite, même chez les enfants plus âgés souffrant d'ankylose congénitale bilatérale du coude.

## ZUSAMMENFASSUNG

Die Operation der angeborenen Ankylose des Ellbogengelenkes war nicht in der Literatur besprochen als der Verfasser zwei doppelseitige operierte Fälle im Jahre 1946 mitteilte (operiert 1943). Aber auch später hat der Verfasser keine Mitteilungen von operierten Fällen dieses Leidens gefunden.

Diese vier 1943 operierten Ellbogengelenke wiesen hinsichtlich des linken Gelenkes ein negatives Ergebnis mit erneuter Versteifung auf. Das rechte Ellbogengelenk hat bei beiden Patienten eine begrenzte Beweglichkeit von 30° behalten. Alle Patienten wurden im Säuglingsalter operiert.

Der dritte Fall, den der Verfasser im Jahre 1950 operiert hat und der hier besprochen wird, hat ein gutes, dauerndes und für den Patienten sehr nützliches Resultat ergeben. Das rechte Ellbogengelenk wurde bei diesem Patienten operiert da sie 4½ Monate, das linke als sie 12 Monate alt war. Das rechte musste 3 mal reoperiert werden, zuletzt da sie 8 Jahre alt war. Das Ergebnis nach 6 beziehungsweise 13 Jahren wird mittels Abbildungen und Röntgenbildern beschrieben und demonstriert.

Der Verfasser bespricht die Frage ob man im Säuglingsalter wo die angenommene Inaktivitetsatrophie der vorhandenen Muskeln noch nicht so fortgeschritten ist, operieren soll. Oder ob man bis zum Kindesalter warten kann, da die technische Ausführung der Operation leichter ist und die Nachbehandlungsumstände günstiger anliegen.

Was die Inaktivitetsatrophie anbetrifft, meint der Verfasser nach seinen Erfahrungen mit dem letzten Falle, dass sie augenscheinlich in den für die Streck- und Beugefähigkeit des Ellbogengelenkes so wichtigen dreigelenkigen Musklen nicht so rasch zunimmt. Das Ergebnis mit diesem 8 Jahre alten-allerdings dreimal operierten rechten Ellbogengelenkscheint zu zeigen, dass es möglicherweise günstiger ist in diesem Alter zu operieren.

In jedem Falle soll man die operative Behandlung der doppelseitigen ankylosis cubiti congenita selbst im vorgeschrittenen Kindesalter nicht als hoffnungslos abweisen.

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