FROM THE HOSPITAL OF ÅLESUND.
MEDICAL SUPERINTENDENT: E. HJORT

CONGENITAL ANKYLOSIS OF THE ELBOW-JOINT

BY

H. FROSTAD

The congenital ankylosis of the elbow-joint is extremely rare, and so far only 24 cases have been reported in the literature. Therefore, I think, the 5 cases which I have had the opportunity to study might possibly be of some interest.

The first case was that of a man, 72 years old, who was admitted to the hospital suffering from retentio urinae. On further examination we found that he had a complete ankylosis of both elbow-joints in about a right-angled position. He told that his arms had been thus since he was born, and that he had one sister and one brother with the same anomaly. They were 6 sisters and brothers altogether. On examination of the rest of the family we did not find similar cases, or cases belonging to the same class of abnormities aplasia.

It now appeared that in the neighbourhood of the first family was another one, where two children of seven had the same abnormities. No direct relation between these two families could be shown. The father, however, told a strange story which he meant might explain why his two children had got it. During the pregnancy with the oldest of the children who had anomaly, his wife had gone about imitating the above-mentioned older man. As a supernatural punishment their children had therefore been shaped similarly. Still it was very strange that both the families lived in the same locality, had almost daily intercourse, and that the abnomities in both were precisely the same.

1. A man, aged 72, of medium height. Congenital right-angled ankylosis of both elbow-joints. A fisherman in his youth, later

he was apprenticed to tailoring, and was a tailor til he was 67. He stated that he had always been able to do his work to everybody's satisfaction. Formerly, he had been healthy, apart from

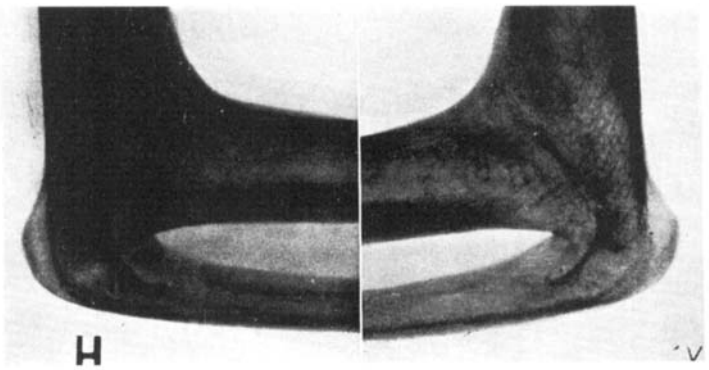

Fig. 1.

the above-mentioned retentio urinae and operation for adenoma of the prostate, for which he had been operated.

2. Woman, aged 64, sister to the first-mentioned patient; little under medium height. Congenital right-angled ankylosis

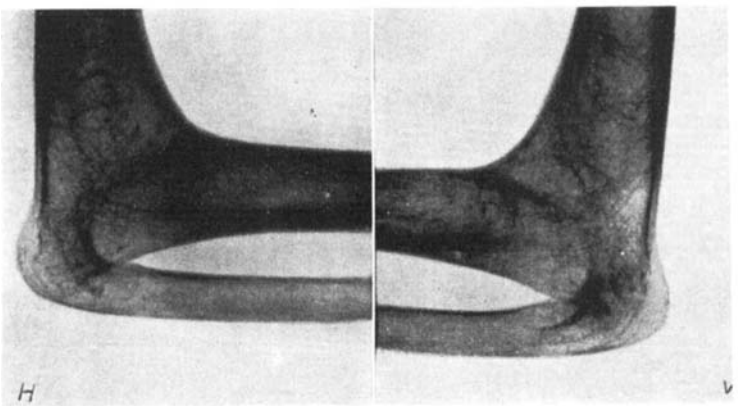

Fig. 2.

of both elbow-joints. While younger she was a maid in the country, doing different sorts of work (dairy—household—and

open air work). She stated, however, that she could do all her work satisfactorily. At the aged of 40, she had an inflammation in her right hand, probably tendovaginitis, with arthritis of some

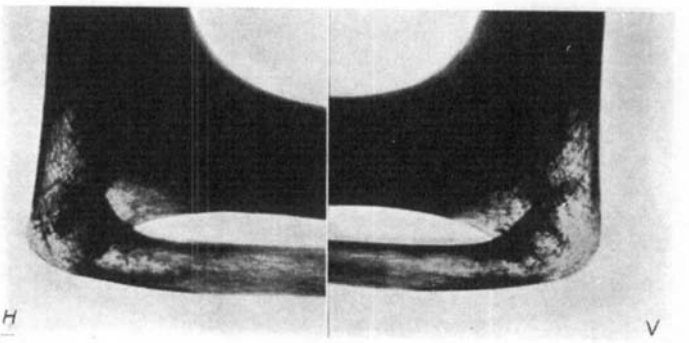

Fig. 3.

finger-joints and the wrist. Later she has only stayed at home and has been house-keeper for an unmarried brother.

3. A man, aged 73, brother of the two above-mentioned of medium height, slim. Congenital right-angled ankylosis of both

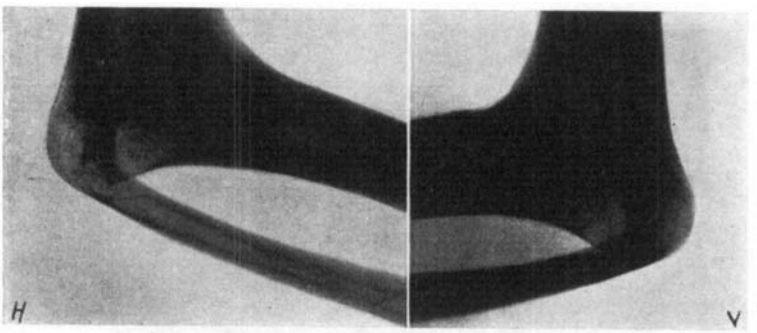

Fig. 4.

elbow-joints. He has been a farmer and fisherman all his life. He has always been healthy, and he still does farm-work in summer. In winter he makes and mends his nets. He has done his work perfectly.

4. A youth, aged 19; slightly under medium height. Congenital ankylosis of both elbow-joints, in a right-angled position on the left arm, and an angle of 95 degrees on the right. He works partly as a farmer, partly as an upholsterer in his fathers workshop. He does farmwork very well indeed, but as an upholsterer he cannot work so quickly as the others.

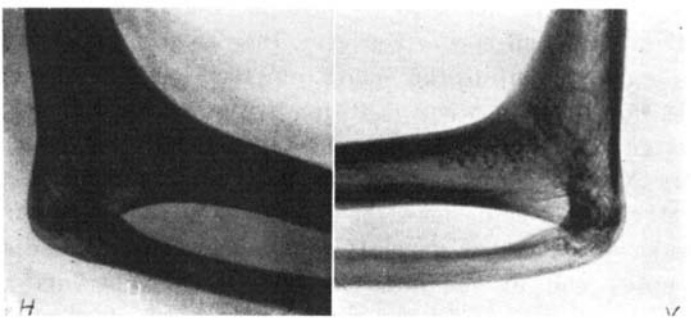

Fig. 5.

5. A girl, aged 12, sister of the 19 years old boy, and next to him in the family, of medium height, slim. Congenital ankylosis of both elbow-joints, at an angle of 95 degrees on the left arm, and 105 degrees on the right. She is still goes to school, and does excellently in the various subjects, also in writing, drawing and needle-work. Her arms are rather plump, which may be due to the fact that there is still with her an infantile distribution of the subcutanous fat. The muscles are in a similar condition to the other cases. On the second, third and fourth fingers of the right hand small nodules have developped on the flexure-sinews, which possibly may be due to unaccustomed use.

Certain things are common in all five cases. They have not, of course, been able to do work neccesitating the bending of the elbow-joints. They have not been able to touch the head, neck or upper part of the trunk above the level of the xiphoid process in front and the 10.—12. dorsal vertebrae posteriorly. They have not been able to tie their ties or to butten up their clothing across the chest. If not assisted by others, they have

only managed to wash their head, neck and upper part of the trunk by means of a longhandled brush. The men have not been able to shave themselves. The 64-year-old woman says she has not been able to bake bread, as constant bending and stretching of the elbow-joints are necessary in kneading the dough. To eat has been difficult for all of them, both on account of the ankylosis of the elbow-joints and of the pronated position of the hand. They have managed by bending the head forwards to meet the fork and spoon. However, they have not been able to raise a glass or cup to the mouth, nor themselves to clean their teeth. For them to write is difficult, as the hand is strongly pronated and conseqnetly the volar surface rests on the desk.

By X-ray examination of the arm, we found the scapula and clavicule to be normal; the size of the upper and lower-arm to be somewhat below normal; and the cavitas glenoidalis and the upper end of the humerus with their tubercula to be normally developed. In the elbow region the humerus continues, without interruption by corticalis and the marrow cavity to the radius, which has the same thickness as the humerus. The radius tapers quickly to the hand. On fig. 2, right side, we can see a rather fine capillary fissure at the junction of the humerus and the radius, but otherwise the bone structure continues without interruption. In all the cases condylus med. humeri is well developed, and the ulna is directly connected with it. On fig. 3, right side one may see a suggestion of a joint fissure between the condyllus med. humeri and ulna. Otherwise we see no suggestion of a joint fissure and the corticalis and the bone structure continues uninterrupted from one bone to another. On fig. 1 we see that the upper end of the ulna engages in a bifurcated way round the condyle, and there is a suggestion of a formation of condylus lat. humeri. On fig. 1 and 3 there is a suggestion of an olecranon. In the upper part of the lower arm the ulna is considerably thinner than usual, whilst the radius is of the same thickness as the distal end of the humerus. Both bones taper towards the carpus, but particularly so the radius. The articular surfaces facing the carpal bones are normal and the stylioid processes of the ulna and radius are present. Both,

however, are poorly developed. The carpal and metacarpal bones and the phalanges of the fingers are normal in number and appearance, apart from the arthritic changes in No. 2.

The general examination of these 5 cases shows, with small deviations, similar conditions in all of them. The whole upper extremity appears to be somewhat shorter than usual. The length of the arm measured from the point of the acromion to the very top of the angle in the elbow-region and further to the wrist may be seen from following table.

	R.upper arm length.	R.lower arm length.	L.upper arm length.	L.lower arm length.
1.	29 cm.	23 cm.	29 cm.	22 cm.
2.	25 cm.	20 cm.	25 cm.	20 cm.
3.	28 cm.	23 cm.	26 cm.	21 cm.
4.	29 cm.	20 cm.	29 cm.	20 cm.
5.	20 cm.	16 cm.	20 cm.	16 cm.

The muscles of the shoulder are strongly developed, particularly the deltoid, which may easily be seen down to its insertion on the upper arm, whereas the rest of the upper and lower arm is very thin and covered with flabby tissue. The circumferences of the upper arm, just below the insertion of the deltoid varied between 17 and 15 c. The circumference of the lower arm, 8 cm. distally to the ankylosis, varied from 19 to 16 cm.

On the upper arm the thin muscle-belly belonging to the caput longum bicipitis might be made out indistinctly by abduction of the arm under pressure. In the same way, a little belly of the caput longum tricipitis, may, by adduction, be palpated on the upper part of the upper arm. Their essential function, however, is bending and stretching of the elbow and consequently they are completely atrophic. Musculus brachialis cannot be palpated.

On the lower arm, m.m. pronator teres, pronator quadratus, brachioradialis, anconeus, supinator and extensores carpi radiales, are nearly completely atrophic. The other moving muscles

in the wrist, which stretch and bend the fingers, are almost normally developed. But the lower arm gets a particular, sausage-formed appearance. The muscles of the hand are strong and normally developed. The movements of the shoulder-joints are normal. The lower arm is entirely pronated so that the flat vola manus rests on the table, the arm resting on the latter. Pronation and supination are nullified. The wrist and the fingers move normally. In the elbow region a projection corresponding to condylus medialis humeri, to which the upper end of the ulna is fixed, may be palpated. On the other hand neither condylus lateralis humeri nor alecranon may be palpated.

In the Scandinavian literature a case of a similar kind is mentioned previously by Ragnar Romanus, who has collected 23 cases reported by other authors. The ankylosis between the bones of the upper and lower arm they had in common. The angle varied from 180 degrees, outstretched position to on acute angle. Most of those indicated lay between 120 and 150 degrees. In all the humerus continues, without interruption by the corticalis and the marrow cavity, to the radius, or, if the ulna is completely missing, to a bone of the same thicknes as humerus. In one, having bilateral ankylosis a small fissure in the angle might be seen, which suggested a division between the 2 bones. In most of them the upper arm was shorter than usual. In the proximal end of the upper arm caput humeri was as a rule, small and deformed, and in some of them both tuberculum majus and minus were missing.

In some both the radius and ulna are present, and in the upper end they are ankylotically connected with humerus as the principal bone. In some cases the radius and the ulna had grown together in the upper part. With some the ulna was completely missing, or there was a small rudiment left at the upper and lower ends, without any connection between them. In some there was only a small rudiment left at the upper end. Where ulna was completely missing or was defective at the lower part, the 4th and 5th metacarpal bones together with the corresponding fingers and one or two of the ulnary carpal bones the were always missing. The metacarpal and carpal bones,

however, varied greatly in number and shape, and in one case the carpal bones had grown together into a flat bone, but with a suggestion of lines of division corresponding to the normal gaps of joints. Only in 4 cases was the ankylosis bilateral.

The most astonishing fact, on comparison of the 5 mentioned cases with the others reported, is that all cases are almost quite similar. The upper arms are normal in the proximal part, and the deformity is bilateral in all of the. Ulna is present in its full length, with normal joints of the hand and wrist.

To be able to understand some of the causal links of this anomaly some features of the development of the skeleton of the upper extremity will be mentioned. The germs of the upper extremity develop in the 3—4 fetal week like a small bud on on the Wolffion lists or the lists of the extremity, which stretch along the whole body. At first they are coated with a single layer of epithelum. The germs are situated on a level with the 4 lowest cervical- and the uppermost thoracic-myotoms. In the 3rd. week the bud is filled with mesenchyme rich in vessels, which is supposed to originate from the parietal leaf of the unsegmented mesoblast. At the end of the 4th week one may see that an axial, strongly densified, region of tissue is being formed in this mesenchyme, the first beginning of the skeleton of the upper extremity. So it has got the name of scleroblastem.

The densification appears first in the middle, at the base of the germs, corresponding to the upper end of the humerus. From this point it grows rapidly in proximal and distal directions. One may already here make out the different parts of the skeleton but indistinctly. During the further development one may distinguish between a broader, proximal part which is the shoulder, and a somewhat smaller, distal one which is the palm of the hand, formed like a leaf, which represents the free extremity. In the middle of the distal part a bend, being the future elbow, is formed.

In the fifth fetal week corresponding to a 11 mm. long embyo a pre-cartilage is formed corresponding to the central part of the bones to be, and then one can already discern

the foundation of the humerus, ulna and radius. This skeleton formation begins to chondrify during the sixth week, and the process is finished at the end of the seventh week. The pericondral bone formation begins at the humerus, ulna and radius at the end of the second month, while the enchondral centres of ossification in the epiphysis first appear after birth.

The development of the joints is particularly important to understand the deformities. Even if the skeleton formation in the pre-cartilage state forms a continuous whole, the scleroblastem arises, however, generally at the places where the joints are to be, somewhat undeveloped, and does not really chondrify, and it changes to the pre-stages of the pericondrium. While the chondrified parts grow in length, this jelly-like medium tissue between them grows continually thinner, and undergoes partial athrophy, by which a joint fissure arises in its place. The circumferential part of the medium tissue is changed into accessory ligaments and capsula fibrosa, the inner face of which is at last coated with a layer of endothelial-like cells, synovialis. By the ossification of the epiphysis a part of the cartilage is left and forms the cartilage of the joint. The cavity of the elbow joint begins to shape in the third fetal month.

In some cases it is evident that the cause of this anomaly is due to a hereditary condition. Thus Siwon in 1928 mentioned the case of a four-year-old girl with congenital ankylosis of both elbow joints at an angle of 150 degrees. Her mother had a bilateral ankylosis of both elbow joints at an angle of 135 degrees. The mother again on her father's side had an aunt and a grandmother with similar anomalies. Likewise Joachimsthal in 1900 tells about an elderly man and his thirteen-year-old daughter who both had congenital ankylosis of both elbow joints. The five cases which I have mentioned appear to support the theory of hereditary genesis.

The changes which lead up to the congenital ankylosis in the elbow joints were first to appear at the formation of the scleroblastem, and at the formation of the pre-cartilage and the cartilage. One must suppose that the scleroblastem does not show

any immature state corresponding to the elbow joint. When the chondrification starts in the humerus, it also continues, completely, to the spot corresponding to the joint, it is not here an object to either change or athrophy, and the formation of the joint cavity does not take place.

Some authors have thought that the cause should be amniotic changes just like the formation of fibres and intergrowths. However, the changes which lead up to the congenital ankylosis must occur so early that the amniotic changes which may appear, are of no importance. Even if the foundation of the arm was fixed by fibres, this would have no influence upon the chondrification and the formation of the joint cavity. If amniotic changes played any important part, one might expect to find congenital ankylosis of several of the other large joints. It must be a characteristic, rare congenital, anomaly which is limited to the elbow joints.

The possibility of operative formation of a new joint has been discussed, but never attempted. It would be rather useles to try, one would think, particularly because the muscles which should move in the joints are completely athrophic.

RÉSUMÉ

Il est rendu compte de 5 cas d'ankylose congénitale dans l'articulation du coude. Tous ces cas sont bilatéraux et à peu près similaires. Trois d'entre eux ont été observés dans une famille et deux dans une autre famille. Ces circonstances appuient la théorie de la genèse héréditaire. Il est fait mention également de certains faits concernant le développement des extrémités supérieures et on relève notamment que les modifications qui conduisent à l'ankylose apparaissent très tôt, déjà au moment de la formation des ostéoblastes et de l'ossification enchondrale.

SUMMARY

5 cases of congenital ankylosis in the elbow joint are mentioned. All are bilateral and almost quite similar. 3 of them occurred in one family, and the 2 others in another family. These facts support the theory of hereditary genesis. Some features of the development of the upper extremity are mentioned, and it is pointed out that the changes which lead up to ankylosis must appear early, during the formation of the scleroblastem and the chondrification of the germs of the bones.

ZUSAMMENFASSUNG

Es wird über 5 Fälle von kongenitaler Ankylose im Ellbogengelenk berichtet. Alle sind bilateral und beinahe ganz gleichartig. 3 der Fälle kamen in einer Familie vor und die beiden anderen in einer anderen Familie. Dieser Umstand stützt die Theorie der erblichen Genese. Gleichzeitig werden einige Eigentümlichkeiten in der Entwicklung der oberen Extremitäten erwähnt, und es wird darauf hingewiesen, dass die Veränderungen, die zu der Ankylose führen, schon frühzeitig auftreten zur Zeit der Ausbildung des Skleroblastems und der Ossifikation der Knochenkerne.

LITERATURE

Gb. B. Gruber: Die Morphologie der Missbildungen des Menschen und der Tiere. 1937. p. 300.

R. Barden: In Keibel und Malls Handbuch der Entwicklungsgeschichte des Menschen. 1910. p. 368.

H. K. Corning: Lehrbuch der Entwicklungsgeschichte des Menschen. 1921. p. 204.

Ragnar Romanus: Acta Ortopedica Scandinavica 1933. p. 291.

A. Herzog: Fortschritte a. d. Gebiete d. Röntgenstrahlen. Vol. 36. p. 407.