CONGENITAL PSEUDARTHROSIS OF THE CLAVICLE

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Congenital pseudarthrosis of the clavicle is a rare condition, first reported by Fitzwilliams in 1910. Alldred (1963) reported nine cases and distinguished it from cleido-cranial dysostosis and birth fracture of the clavicle. Other reported cases have brought the total to nineteen (Saint-Pierre 1930, Carpenter and Garrett 1960, Marmor 1966, Caffey 1967, Jinkins 1969). We report here another thirteen patients with this problem, describe others with allied conditions, present some patients with a strong family incidence and report on some relevant embryological studies.

**TABLE I**

**PATIENTS WITH CLASSICAL CONGENITAL PSEUDARTHROSIS OF THE CLAVICLE**

<table>
<thead>
<tr>
<th>Case number</th>
<th>Side</th>
<th>Age at presentation</th>
<th>Obvious lump</th>
<th>Injury</th>
<th>Pain</th>
<th>Family history</th>
<th>Treatment</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Right</td>
<td>1 day</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>None</td>
<td>Seen 3 months later. No union</td>
</tr>
<tr>
<td>2</td>
<td>Right</td>
<td>4½ months</td>
<td>Yes</td>
<td>No</td>
<td>Discomfort from sixth year</td>
<td>No</td>
<td>At 7 years repaired with Kirschner wire and bone graft. Resection of lesion</td>
<td>Union</td>
</tr>
<tr>
<td>3</td>
<td>Right</td>
<td>11 months</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Right</td>
<td>13 months</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Repaired with Kirschner wire and graft at age of 3</td>
<td>Uneventful union</td>
</tr>
<tr>
<td>5</td>
<td>Right</td>
<td>22 months</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Right</td>
<td>2 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Repaired with threaded wire and graft</td>
<td>Wire split proximal fragment of clavicle. Despite this, union was obtained</td>
</tr>
<tr>
<td>7</td>
<td>Right</td>
<td>3 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Right</td>
<td>3½ years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Compression plate and graft at 16 years</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Right</td>
<td>5 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Repaired with Kirschner wire and graft. Lesion excised</td>
<td>Union</td>
</tr>
<tr>
<td>10</td>
<td>Bilateral</td>
<td>7 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>None</td>
<td>Anoxia at birth following accidental haemorrhage</td>
</tr>
<tr>
<td>11</td>
<td>Right</td>
<td>7 years</td>
<td>Yes</td>
<td>No</td>
<td>Numbness and tingling in arm</td>
<td>No</td>
<td>Repaired with threaded wire and bone graft at age of 15</td>
<td>Union</td>
</tr>
<tr>
<td>12</td>
<td>Bilateral</td>
<td>8 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Repaired with Kirschner wire and graft</td>
<td>Union</td>
</tr>
<tr>
<td>13</td>
<td>Left</td>
<td>19 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>None</td>
<td>Pseudarthrosis was an incidental finding. Dextrocardia present</td>
</tr>
</tbody>
</table>
Case 1. Figure 1—Radiograph on the first day of life taken for other reasons. Congenital pseudarthrosis of the right clavicle can be seen. Figure 2—Three months later there was no sign of union.

Case 5—Radiograph of a child of 22 months taken when the parents noticed a lump in the right shoulder.

Case 8—Radiograph at 16 years of age showing classical pseudarthrosis of the right clavicle.
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Case 9. Figure 5—Photograph taken at the age of 5, when the child presented with a lump that distressed the parents. Figure 6—Radiograph taken when the patient was first seen shows pseudarthrosis. Figure 7—A lordotic view shows the wide separation of bone fragments. Figure 8—Three months after bone grafting solid union had been obtained.

Case 9—Photomicrograph of the operative specimen shows sclerosis of subchondral bone and a well formed cartilage cap at the site of the pseudarthrosis. (Haematoxylin and eosin, $\times 45$.)
The patients with classical congenital pseudarthrosis of the clavicle presented soon after birth, or in childhood, with a painless lump on the clavicle, without any history of injury, pain or major disability. The deformity tended to become more conspicuous as the child grew, and consisted of a false joint between enlarged ends of the clavicle just lateral to its mid-point. The sternal fragment lay in front of and above the acromial fragment. Clinical details of the

![Fig. 10](image1)

![Fig. 11](image2)

![Fig. 12](image3)

![Fig. 13](image4)

Case 10. Figure 10—Photograph of a boy aged 8 years in whom bilateral congenital pseudarthrosis was an incidental finding. There was no marked clinical deformity. Figure 11—Hypermobility at the sites of pseudarthrosis is revealed. Figure 12—Radiograph at the time of presentation reveals bilateral pseudarthrosis of the clavicle. Figure 13—Eight years later the radiographic appearance is unchanged.

patients are summarised in Table I (Figs. 1 to 16). In two cases in which histological material was obtained the bone ends were sclerotic and covered with a thick cartilage cap (Fig. 9). In ten patients the pseudarthrosis was on the right side, in two it was on both sides and in one case it was on the left side. It is noteworthy that this last patient also had dextrocardia.

A second group of patients with pseudarthrosis of the clavicle from varying causes was seen. The clinical details are summarised in Table II and Figures 17 to 24. The one fracture
that occurred at birth (Case 14) healed readily, suggesting that congenital pseudarthroses are not missed birth fractures. The child of four (Case 15) had repeated injuries. Operative repair was attempted unsuccessfully and nine years later a persistent painful lump was excised.

Case 11. Figure 14—Radiograph of the right clavicle of a 15-year-old boy who presented for removal of the bony lump. Figure 15—Radiograph after operative fixation of the clavicle with a Kirschner wire to show bone union.

Case 13—Radiograph of a 19-year-old boy who had no symptoms from the left clavicular pseudarthrosis. Dextrocardia was found to be present.

The lesion could not have been a congenital pseudarthrosis and it did not look at all like one (Figs. 17 and 18). There was a fibrous connection between the bone ends, but no cartilage. The girl with cleido-cranial dysostosis, seen for flat feet, was not concerned with her shoulders,
TABLE II
MISCELLANEOUS PSEUDARTHROSIS OF THE CLAVICLE

<table>
<thead>
<tr>
<th>Case number</th>
<th>Side</th>
<th>Age at diagnosis</th>
<th>Obvious lump</th>
<th>Injury</th>
<th>Pain</th>
<th>Family history of fractured clavicle</th>
<th>Treatment</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>14</td>
<td>Right</td>
<td>1 day</td>
<td>Yes</td>
<td>Difficult delivery</td>
<td>Tender lumps No</td>
<td>None</td>
<td>Healing with abundant callus in 2 weeks</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>Left</td>
<td>4½ years</td>
<td>Yes</td>
<td>Three times in one year</td>
<td>Yes No</td>
<td>Attempted repair at 4 years. Resection of painful persistent pseudarthrosis at 13 years</td>
<td>At 16 years good painless movement was present</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>Bilateral</td>
<td>17 years</td>
<td>Not very obvious</td>
<td>No</td>
<td>No No No No</td>
<td>None</td>
<td>Known cleido-cranial dysostosis</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>Right</td>
<td>14 years</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Resected and fixed with wire; then grafted with union</td>
<td>Seven injuries followed by pain since age of 3 years</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>Right</td>
<td>16 years</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Resected and fixed with wire; then grafted with union</td>
<td>Six injuries with pain since age of 2 years. Broke left clavicle once but it united</td>
<td></td>
</tr>
</tbody>
</table>

Case 15. Figure 17—Radiograph of the left clavicle of a 3-year-old child showing a fresh fracture. Figure 18—Radiograph taken one year later, after the clavicle had fractured twice more, shows established pseudarthrosis. Figure 19—Radiograph after excision of the pseudarthrosis showing the clavicular fragments fixed with a Kirschner wire. Figure 20—Bone union did not occur and the wire was removed.
Case 15. Figure 21—The patient continued to have pain from this pseudarthrosis; at the age of 13 wide excision of the bone ends was performed. The radiograph shows the final result, when the patient was free of symptoms. Figure 22—Photomicrograph of the operative specimen shows sclerosis of the bone ends with a mass of fibrous tissue between. There has been no formation of cartilage. (Haematoxylin and eosin, × 4.)

Case 16. Figure 23—Photograph of a 17-year-old girl with cleido-cranial dysostosis. There is no conspicuous deformity although a lump is seen on the left side. The patient suffered no symptoms. Figure 24—Radiograph in the same case. It will be seen that the bone ends are tapered and not bulbous.
where the deformity was inconspicuous (Fig. 23). Radiographs showed well separated tapering ends of the clavicular fragments, quite different from those seen in congenital pseudarthrosis (Fig. 24).

The presence of such an unusual condition as traumatic pseudarthrosis of the clavicle in childhood in two boys in one family suggests that a congenital abnormality was present before injury (Cases 17 and 18). This view is supported by the presence of cartilage and a false joint at the pseudarthrosis.

The third group of patients with pseudarthrosis of the clavicle gave a family history of the condition. Two families were involved (Table III). The first patient (Case 19) said that

<table>
<thead>
<tr>
<th>Case number</th>
<th>Side</th>
<th>Age when first seen</th>
<th>Lump</th>
<th>Injury</th>
<th>Pain</th>
<th>Family history</th>
<th>Treatment</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>19</td>
<td>Right</td>
<td>33 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Aunt and grandfather affected</td>
<td>None</td>
<td>Patient complains of fatigue of left arm unless supported when she sits</td>
</tr>
<tr>
<td>20</td>
<td>Bilateral</td>
<td>50 years</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Rib graft to both sides at age of 21 years, Right side fused</td>
<td>Patient cannot throw with right hand and has to support right arm when sleeping on left</td>
</tr>
<tr>
<td>21</td>
<td>Right</td>
<td>40 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>None</td>
<td>Patient feels cracking in bones, Right arm tires easily; left handed</td>
</tr>
<tr>
<td>22</td>
<td>Right</td>
<td>13 years</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>None</td>
<td>Patient cannot sleep on shoulders, No other disability; was not aware of this until she was 32 years old</td>
</tr>
<tr>
<td>23</td>
<td>Bilateral</td>
<td>37 years</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>None</td>
<td>Bilateral cancellous bone grafts at age of 8 Union with no symptoms</td>
</tr>
<tr>
<td>24</td>
<td>Bilateral</td>
<td>11 years</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Bilateral cancellous bone grafts at age of 5</td>
<td>Successful</td>
</tr>
<tr>
<td>25</td>
<td>Bilateral</td>
<td>5 years</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Unsuccessful surgery at age of 19</td>
<td></td>
</tr>
<tr>
<td>26</td>
<td>Right</td>
<td>37 years</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>None</td>
<td>Right arm becomes numb if she sleeps on it</td>
</tr>
<tr>
<td>27</td>
<td>Right</td>
<td>21 years</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>None</td>
<td></td>
</tr>
</tbody>
</table>

her aunt and grandfather had right clavicles just like hers, but no other abnormalities. She did not regard her abnormal shoulder as a handicap.

The other patients with congenital pseudarthrosis of the clavicle were all members of one family (Fig. 26). They were all of short stature and several of them had a high palatal arch and irregular upper dentition. No other skeletal abnormality was found in the eight members personally examined. Clinical details of these patients are recorded in Table III. From the family tree it seems likely that in this family congenital pseudarthrosis of the clavicle represents a genetically determined defect with an autosomal dominant type of transmission. In generation II the eldest brother’s condition was unknown, the second brother was definitely involved bilaterally according to his family, but it was not known whether the rest of the family were affected.

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The members of the recorded third and fourth generations had gathered together on one occasion for a family reunion and their family peculiarity had been much discussed; since the deformity was easily recognisable, the information they gave was probably reliable. However, only those marked X were personally examined.

**EMBRYOLOGY OF THE HUMAN CLAVICLE**

We studied the development of the clavicle in human embryos. Embryos of 12, 16, 17, 18, 19 and 20 millimetres were examined in serial sections, the age assessment from crown-rump measurement being corroborated in each case by studying the stage of development of the heart. Under the dissecting microscope clavicles were removed from 32, 41, 58, 90, 170, 200 and 270 millimetre embryos. These clavicles were sectioned longitudinally and studied histologically.

We were unable to identify the clavicle in the 12-millimetre embryo. In the 16-millimetre embryo (Fig. 27) there was a definite condensation of mesenchymal tissue radiating towards the sternum from the anlage of the acromion. In the 17-millimetre specimen this condensation was easily seen (Fig. 28). In the 18-millimetre embryo (Fig. 29) the clavicle was easily seen, and in a higher power view (Fig. 30) from the same slide it can be seen that the undifferentiated mesenchymal tissue has become "precartilage" or "honeycomb cartilage". The section in Figure 29 shows a continuous single anlage for the clavicle. A deeper cut from the same embryo (Fig. 31) appears to show two centres for the clavicular anlage. This appearance has been interpreted in the past as meaning that the clavicle has two centres of ossification. Our study does not support this opinion, for whenever this appearance was seen, a connection between the two "centres" could be found in adjacent cuts.

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**Fig. 25**
Case 21—Radiograph of the right shoulder of a 41-year-old man who had only minimal symptoms from this pseudarthrosis.

**Fig. 26**
Cases 20 to 27—Pedigree of family with congenital pseudarthrosis of clavicles. Although this family tree is based on the examination of only eight members, the history of the family as it affects generations III and IV is reliable because the deformity is so easily recognised. It is not certain whether the eldest brother in generation II was affected, nor is it certain whether the eldest brother in the second family in generation III was affected. The family thought he was not. (The numbers correspond to case numbers in Table III.)
Section of the 19-millimetre embryo showed further development of the rather open-looking honeycomb cartilage. Those from the 20-millimetre embryo showed well differentiated precartilage clavicles (Fig. 32), and with greater magnification (Fig. 33) early osteoid deposition under the enveloping membrane and in the centre of the shaft could be seen.

The longitudinal sections from the larger embryos (Figs. 34 to 41) showed a steady progression of the deposition of osteoid, so that the central part of the shaft became osteoid

(Fig. 34), replacing precartilage except at the ends, where the precartilage changed to hyaline cartilage (Fig. 37). Thus the central part of the shaft was formed directly from the precartilage anlage and the ends by endochondral ossification.

We conclude that the human clavicle develops in the following stages: 1) mesenchymal condensation; 2) a change in the mesenchyme to form precartilage, which grows rapidly; 3) the precartilage forms a complete and well defined model of the clavicle, enveloped in a surrounding membrane; 4) deep to the enveloping membrane and in the centre of the shaft osteoid appears
In this photomicrograph of the 18-millimetre embryo the clavicular anlage appears to have two centres but comparison with Figure 30 which was taken from the same specimen shows that it is because the section was taken through a curved structure. (Haematoxylin and eosin, × 38.)

Photomicrograph of a 20-millimetre embryo. The left clavicle is easily seen. (Haematoxylin and eosin, × 6.)
and spreads toward the ends of the bone, replacing precartilage; 5) at the ends of the bone hyaline cartilage replaces the precartilage, and osteoid gradually extends into these cartilage end-caps.

**DISCUSSION**

Our patients with classical congenital pseudarthrosis of the clavicle all had conspicuous lumps on the shoulder, with excess movement, but without any history of injury, pain or serious disability. Some, however, had minor complaints, and either they or their parents wished to be rid of the lump and to obtain a stronger shoulder. Only in such circumstances should treatment be offered. Any form of simple fixation with cancello-cortical onlay grafts is adequate to promote solid union. The dangers of operation are related to the proximity of the subclavian artery (Case 15), splintering of the bone if the operation is done at a very early age (Case 6) and infection. It is probably wise to defer operation until pre-school age, as it is then technically easy.

The treatment of these patients is optional and, when desired, easy. The lesion remains difficult to understand. Why should all the pseudarthroses, if unilateral, be right-sided, or at least on the side opposite to the heart? Patients with a congenital lesion are different from those with an acquired one in their freedom from pain and in the histological picture. They differ from patients with cleido-cranial dysostosis in that the lump is large and the bone ends are close together. In the family with large numbers of these lesions some members had inconspicuous lumps and greater defects of bone at the site of the lesion, more like cleido-cranial dysostosis. It is possible that in these patients the lesions did not have the same cause. Some may in fact result from unremembered injuries, and others may represent a "forme fruste" of cleido-cranial dysostosis; yet others may be a specific entity related to an acquired or genetically determined defect in the development of the clavicle.
Figure 34—Photomicrograph of the dissected clavicle of a 32-millimetre embryo. Osteoid is spreading from the centre of the shaft towards both ends. (Haematoxylin and eosin, × 25.)

Figure 35—In a 41-millimetre embryo, the amount of osteoid is increasing. (Haematoxylin and eosin, × 25.)

Figure 36—In a 58-millimetre embryo the clavicle is largely osteoid. (Haematoxylin and eosin, × 25.)

Figure 37—In a 90-millimetre embryo only the ends of the clavicle remain cartilaginous. (Haematoxylin and eosin, × 17.)
Figure 38—High power photomicrograph of the clavicle in the 90-millimetre embryo showing the sternal end which is largely well formed cartilage. (Haematoxylin and eosin, ×6.5.) Figure 39—In a 175-millimetre embryo the clavicle is almost all osteoid. (Haematoxylin and eosin, ×6.5.) Figure 40—In a 200-millimetre embryo growth of the clavicle has occurred by the cartilaginous caps. (Haematoxylin and eosin, ×6.5.) Figure 41—In a 270-millimetre embryo the clavicle is very similar in appearance to the familiar structure present after birth. (Haematoxylin and eosin, ×6.5.)
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One theory as to why congenital pseudarthrosis of the clavicle occurs has been that the clavicle normally ossifies from two centres and that occasionally these fail to fuse. Mall (1906) described ossification from two centres, which he said appeared by the thirty-ninth day and were fused by the forty-fifth day. Fawcett (1913) described two ossification centres appearing in two precartilaginous masses in 17-millimetre embryos and uniting in 18- and 19-millimetre embryos. His views, supported by interpretative drawings, have been extensively quoted in the literature, including Alldred's (1963) classical review of congenital pseudarthrosis of the clavicle. Zawisch (1952) described an endodermal osteoblastic stage, a pseudo-chondroblastic stage, and a chondroblastic stage of clavicular development, and supported the concept of two centres of ossification. Koch (1960) presented the best review available of the embryology of the clavicle. He supported the concept of a single centre of ossification, starting in the middle of the clavicle and forming a tube of bone around a honeycomb type of cartilage that spreads towards the ends; the ends continue to grow within normal hyaline cartilage and ossify eventually by endochondral ossification. Our work supports his view. We think that if the clavicle ossifies from two centres this would be abnormal in itself and, while it may happen in cases of congenital pseudarthrosis of the clavicle, it then requires an explanation.

SUMMARY AND CONCLUSIONS

1. Thirteen cases of congenital pseudarthrosis of the clavicle conforming to the classical picture are presented, together with five cases of pseudarthrosis occurring for various other reasons, and nine cases of congenital pseudarthrosis that form a special group because there was a strong family history.

2. Treatment, if desired by the patient, is easy, but should be deferred until the age of four or five years.

3. The etiology remains obscure. Our embryological study does not support the theory that the clavicle normally develops from two ossification centres that may fail to coalesce. Although in thirteen of our patients there was no family history of pseudarthrosis, there was another group of nine patients who presented a strong family history, and it seems certain that in some instances congenital pseudarthrosis of the clavicle can be transmitted genetically.

The authors wish to thank Dr. J. W. A. Duckworth, Professor of Anatomy in the University of Toronto, for his assistance in providing specimens and in helping to interpret the embryology of the clavicle. Only four of these patients were advised or treated personally by the authors, who are indebted to their colleagues at the Hospital for Sick Children, Toronto, and to Drs. John Hazlett, George Kay, Pierre Labelle, Glen MacDonald, Edward Simmons and Michael Simurda for contributing their case records and illustrations to this collection. Drs. Denis Desjardin and James Wiley were especially helpful in making it possible for one author to examine the family (Cases 20 to 27).

REFERENCES


VOL. 52 B, NO. 4, NOVEMBER 1970