CONGENITAL PSEUDARTHROSION OF THE CLAVICLE

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Congenital pseudarthrosis of the clavicle is rare. In the past ten years I have been able to collect nine cases and to trace two more in the literature. The case histories of these children indicate that the condition is not well known, that its natural history has never been adequately recorded and that advice given to parents has often been based on speculation.

The purpose of this paper is to establish that congenital pseudarthrosis of the clavicle exists as a clinical entity, to distinguish it from cranio-cleido-dysostosis and birth fracture of the clavicle, to describe the natural history and to discuss its embryology and treatment.

Cranio-cleido-dysostosis is a familial disorder of the skeleton which may affect many bones, and which is thought to be a disorder of ossification of membrane bones, with most patients having defective or absent clavicles. It is well known that children with this disorder suffer from bossing and other skull deformities, smallness of the facial bones, scoliosis, abnormal epiphyses in the hands and feet and deficiencies of the pelvic ring. Usually there is a family history of bone disorders (Fitzwilliams 1910, Stocks and Barrington 1925, Fairbank 1949).

Congenital pseudarthrosis of the clavicle has been thought to be a manifestation of cranio-cleido-dysostosis, and Fairbank (1949) quoted Stocks and Barrington who described three patients in whom the clavicle was separated into two parts by a pseudarthrosis. In two of these there were other manifestations of cranio-cleido-dysostosis, but in the third, described in great detail by Fitzwilliams (1910), there were no abnormal features other than the pseudarthrosis of the right clavicle. Although this patient was included in Fitzwilliams's series of cranio-cleido-dysostosis, I believe it to be the first carefully recorded case of true congenital pseudarthrosis of the clavicle. The second case was that of a small girl described by Saint-Pierre in 1930 with a pseudarthrosis of the right clavicle, noted shortly after birth, with no history of injury, no other bony abnormalities and with no family history of bone disorders.

Congenital pseudarthrosis has affected the right clavicle in all the eleven patients described. A swelling is found at or soon after birth. Except in Cases 2 and 3, in which the patients were half sisters, there was no family history of bone disorders. None of the children had suffered birth injuries and the lump was always painless. In Case 5 the parents had noticed a reluctance to move the arm soon after birth and inability to push on the arm when crawling. This was attributable to the exceptional mobility of the pseudarthrosis.

Detailed clinical examination of all the children revealed no other bone disorder except in Case 8, in which there was angulation of the right clavicle lateral to the pseudarthrosis.

The situation of the pseudarthrosis appears to be constant, being just lateral to the middle of the clavicle. The bone ends are enlarged, there is a variable degree of painless mobility, and a deformity that increases with age. The relationship of the two fragments is always the same, with the sternal fragment being the larger and lying in front of, and slightly above, the shorter acromial fragment. The reason for this is developmental, and will be described later.

The deformity was accurately described by Fitzwilliams, who wrote of his patient as follows: "The right clavicle was in two portions at the junction of the middle and outer thirds. The sternal end was tilted upwards at its outer extremity so as to override the inner end of..."
the acromial portion and formed such a prominence in the neck as to induce the parents to seek treatment for the child. The acromial portion, owing to the weight of the limb, drooped and swung forwards and inwards as in the ordinary fracture of the clavicle; its inner end lay under the sternal fragment and pointed upwards, backwards and inwards so that the right shoulder was lower, further forward and nearer the mid-line than the left. An asymmetrical appearance was thus given to the upper part of the trunk when seen from in front, while posteriorly the vertebral border of the scapula was more prominent than that of its fellow. The two portions were united by a ligament which became more evident on attempting to separate them. The point of the right shoulder could be forcibly adducted towards the sternum through a considerable distance, the point of discussion acting as a hinge. The prominence

in the neck was not altogether due to the overriding and tilting of the one fragment by the other, but was also accounted for by the fact that their adjacent ends were thickened in a very characteristic manner contrasting with the thin peg-shaped extremities seen in ununited fractures. The deformity usually increases while growth continues and may reach disfiguring proportions (Figs. 2, 5, 6 and 7). The condition is very different from birth fractures of the clavicle with which there is often a history of birth injury. The fracture causes disuse, or pseudo-paralysis of the arm and pain on movement. A lump of callus appears after several days and the radiographic appearances are quite distinctive (Fig. 1).

CASE REPORTS

Case 1—A boy aged two and a quarter years was seen at the Robert Jones and Agnes Hunt Orthopaedic Hospital in 1950 with a lump in the right clavicle. The mother had reported this eleven days after a normal confinement and thought it had been present at birth. There was no family history. The child did not complain and used his arm normally but the deformity had increased with growth. Clinical examination revealed a mobile, painless pseudarthrosis with the medial fragment lying in front of and above the lateral one. Both fontanelles were closed. The teeth, jaws, spine and limbs were normal, as confirmed by radiographs which showed the congenital pseudarthrosis of the right clavicle. He was not treated and no follow-up is available.

Case 2—A girl aged sixteen attended the Timaru Hospital because of a swelling in the right side of the neck which had been present all her life but had increased to such a size that she was very conscious of it. The mother said that the confinement was normal and that she had noticed the lump a few weeks after birth. As it did not appear to cause pain or interfere with arm function she had not sought medical advice. A younger half sister, aged eight, had a similar defect (Case 3). No other cases were known in the family. Examination revealed a large painless lump in the right clavicle, which was in two parts with a mobile pseudarthrosis between. The medial fragment lay in front of and above the
lateral one (Fig. 2). There was asymmetry of the right upper chest and shoulder. Function was good but strength was diminished. At operation the enlarged bone ends were trimmed. A joint cavity was found with articular cartilage on both the bone ends (Fig. 3). The outer end of the medial fragment pointed backwards over a distance of one and a half centimetres. Bone grafting would have been technically very difficult if not impossible. Three years after the operation the deformity was greatly reduced, but there was slight weakness of the shoulder and occasional aching as a result of the operation.

**Case 3**—A girl aged eight years was seen with a small lump in the right clavicle. This was noticed the day after birth and had increased very little in size. She was the younger half-sister of the girl described above (Case 2). Examination showed a mobile pseudarthrosis in the right clavicle with asymmetry of the shoulders (Fig. 4). The skull, teeth, jaws, limb bones and trunk were clinically normal. Radiographs showed a pseudarthrosis of the right clavicle. During the next three years the deformity slowly increased but had not reached such proportions that treatment was needed.

**Case 4**—A boy was found to have a swelling on his right clavicle at birth. Radiographs showed that the clavicle was in two parts, and a birth fracture was diagnosed. From birth to the age of twelve years the swelling gradually increased. Serial radiographs revealed a typical pseudarthrosis of the right clavicle.
clavicle with enlargement of the bone ends (Figs. 5 to 8). He had full shoulder function and could play games normally. Examination of the skull, limb bones and trunk showed no abnormality, and there was no history of birth injury. No other cases were known in the family.

Case 4. Figure 5—Radiograph soon after birth showing the pseudarthrosis of the right clavicle. Figure 6—At the age of four years the deformity is already increasing. Figure 7—At the age of twelve years there is further angulation and enlargement of fragments. Figure 8—This boy shows the typical swelling which is the presenting feature of congenital pseudarthrosis of the clavicle.

Case 5—A girl aged two years three months was seen at the Hospital for Sick Children with a swelling of the right clavicle which had been noticed at the age of six months. She was the third child of West Indian parents. The confinement was normal but the baby had been noticed to use the right arm less
than the left. Shoulder movements caused no discomfort. When she crawled she had difficulty in supporting the trunk with the right arm. The mother had been told that this was a birth fracture and that nothing could be done until the child was five years old. Examination showed a mobile pseudarthrosis of the clavicle with asymmetry of the shoulder. There were no abnormalities of the skull, teeth, jaws, limbs or trunk, and no other members of the family had bone deformities. Radiographs confirmed the pseudarthrosis. At operation, when she was two and a half years old, the bone ends were cut back, thus removing the articular cartilage, and were fixed with an intramedullary Steinmann's pin. Iliac onlay grafts were placed around the gap. Union, confirmed radiologically, quickly occurred (Fig. 9). One year later the clavicle was firmly united and of equal length to the left. The asymmetry of the shoulders had disappeared and function was normal.

**Case 6**—A girl aged two months was seen at the Hospital for Sick Children with a swelling of the right clavicle noticed at birth. She was the first child of parents who gave no family history of deformity. Labour had been induced at forty-one weeks because of hypertension; the delivery was normal. A radiograph, taken on the first day, was reported to show "an old fracture with blunted ends." In fact the appearance was typical of a congenital pseudarthrosis. The child has been seen at intervals. At the age of five and a half the lump had not increased much but the mobile pseudarthrosis was easily felt. The right shoulder was thinner and set more forward, indicating the gradual onset of secondary deformity. Clinical examination of the skull, teeth, jaws and trunk was normal, but both first metatarsals were short. There were no other abnormalities.

**Case 7**—A girl aged six months presented with a lump on the right clavicle, first noticed by her father, a doctor, when the child fell from a chair. He had also found that the swelling could be moved without any discomfort. There had been no previous injury and delivery had been normal. There was no family history. Clinical examination revealed no other skeletal abnormality, and the pseudarthrosis was confirmed by radiographs. At operation at the age of eighteen months the pseudarthrosis was excised until there was bleeding from the bone ends. A Kirschner wire was used to fix the fragments and a tibial graft fixed across the gap and held by encircling wires. The area was packed with chips. Two years later radiographs confirmed that union had occurred (Fig. 10). The wires have since been removed, and the shoulder has become normal in contour and function.
Case 8—A girl aged nine months was brought to the Hospital for Sick Children with a swelling on the right clavicle first noticed at the age of three months. The swelling had steadily increased in size, but was causing no disability. There was no history of injury and delivery had been normal. There was no family history. Clinical examination revealed no abnormality other than an acute angulation of the right clavicle with a pseudarthrosis at its apex. Radiographs confirmed a pseudarthrosis at the usual site but the lateral fragment was angled 90 degrees backwards as it turned towards the acromion. At first sight it seemed as though there had been a fracture at this site but it was undoubtedly compensation for the acute forward angulation. At the age of two years the pseudarthrosis was exposed at operation, revealing the sclerotic bone ends covered with cartilage, which were removed, exposing normal bone. Chips of bone, removed locally, were packed between the bone ends. One year later the defect remained ununited, the pseudarthrosis could easily be felt and the deformity was increasing.

Case 9—A boy aged three was brought to the Hospital for Sick Children with a symptomless swelling of the right clavicle. The mother had had a normal confinement and had first noticed the swelling when the baby was two months old. She had not brought the child earlier as he was free of symptoms and was using the arm normally. Clinical examination showed a pseudarthrosis of the right clavicle, with no other abnormality of bone, and radiographs confirmed the pseudarthrosis. A bone grafting operation was done at the age of five. The ends of the fragments were freshened and a portion of rib was used to bridge the gap. A Kirschner wire was driven across the acromio-clavicular joint and along the medullary cavity of the lateral fragment to transfix the piece of rib and the medial fragment. After eight weeks the wire was removed. Union occurred with a slight step between the medial edge of the graft and the medial fragment.

DISCUSSION

The constant feature of all cases was the site of the pseudarthrosis and the relationship of the two fragments. The histories suggested that the lesion was well established at birth and that it resulted from a failure of normal ossification.

Figure 11—Sagittal section of left clavicle of 17 millimetre (Robinson) embryo showing: 1) the connective tissue jacket; 2 and 2a) the precartilage; and 3) bone. (Sk. = skin; C. Cl. lig. = costo-clavicular ligament; Cor. pr. = coracoid process.) Figure 12—Drawing of a model of the right shoulder girdle of the 17 millimetre Robinson embryo, viewed from behind and showing the relationship between the sternal (S.S.) and the acromial segments (A.S.) of the clavicle. (St. M. = sterno-mastoid muscle; P.M. = pectoralis major muscle; C.M. = cleido-mastoid muscle; T. = trapezius; Ac. pr. = acromial process; C. Cl. lig. = costo-clavicular ligament; C. pr. = coracoid process; Sc. = scapula.)

The normal ossification of the clavicle has been debated by anatomists for many years. Opinions have varied as to whether the clavicle is preformed in membrane or in cartilage, but it is now generally agreed that the latter is correct. Although the development and ossification of the human clavicle, as described by Fawcett in 1913, makes the anatomy of the lesion very clear, it does not throw any light on the cause.
The clavicle can be distinguished in an 11 millimetre embryo and at this stage consists entirely of connective tissue. At the 15 millimetre stage two separate masses of precartilage are seen and at the 17 millimetre stage (between the fifth and seventh weeks) a separate centre of ossification is evident in each mass of precartilage.

Fawcett's description states that at this stage each segment in cross section shows the following parts (Fig. 11): 1) externally a connective tissue jacket of periosteum or perichondrium. 2) A cylinder of precartilage. 3) A central mass of bone.

Both masses of precartilage lie in a common jacket of perichondrium and the bony masses in them are placed at right angles to one another, that is, the medial mass when seen from the front is oval, whereas the lateral mass is oval when seen from above. The outer extremity of the medial mass of precartilage slightly overrides the outer mass lying above and in front of it (Fig. 12).

1. 
2. [Diagram of the right clavicle showing stages of ossification]
3. 4. [continued diagrams]
5. 6. [continued diagrams]

**Fig. 13**
A scheme of the development of the right clavicle, showing stages of ossification. (C.T. = connective tissue; P.C. = precartilage; C. Cl. lig. = costo-clavicular ligament; C. = calcified cartilage; B. = bone; Br. = bridge; O. = ossification; D.T. = dense tissue.)

At a later stage the precartilaginous masses fuse by their adjacent extremities. The fusion line is well seen in several specimens about the 18–19 millimetre stage, when horizontal sections of the clavicle show two bony masses separated by a non-stained precartilaginous interval. In this interval a further advance brings about union by bone between the two independently ossified centres (Fig. 13). The extension takes place in the bridge of precartilaginous tissue which unites the two main masses of precartilage.

Fawcett concludes: "Cases of existence of the clavicle in two apparently separate masses may, I think, be explained on the ground of non-ossification of the precartilaginous bridge connecting the sternal and acromial segments with one another during the 19 millimetre stage. The bony connection is, for a long period, only a slight one."

A study of these cases shows that in no case did union occur without bone grafting. In most, the deformity steadily increased, reaching such proportions that it constituted a cosmetic disability in girls. The natural history is therefore one of increasing deformity of the shoulder and chest but with little functional disability. The rate at which deformity develops is greater when the pseudarthrosis is very lax. In girls the unsightly lump when the shoulders are bared, and the difficulty of keeping shoulder straps on the deformed shoulder warrant consideration of surgical treatment. In boys such problems do not arise and the appearance has to be weighed against the small functional disability and the possibility of failure. The scars have not been a cause of worry to any patient.

The choice of treatment lies between acceptance of the deformity, bone grafting, or reducing the swelling.
From a study of the cases in this series the following plan is suggested. Bone grafting should be done in all children up to the age of eight. The operation is best done between the ages of two and four when the child is big enough and yet has the best chance of union before much deformity develops. Probably the most satisfactory bone grafting technique is to freshen the bone ends, to apply an onlay graft of rib or iliac bone and to fix the fragments internally.

In children over the age of eight the anatomy of the lesion may make bone grafting difficult and acceptance of the deformity is reasonable especially in boys. When the lump is unsightly in older girls trimming of the lump will improve appearance. The disability is unlikely to be sufficient to warrant removal of the whole of one or both fragments.

**SUMMARY**

1. Congenital pseudarthrosis of the clavicle is described.
2. The distinction from cranio-cleido-dysostosis and from birth fracture is stressed.
3. Details are given of two patients reported in the literature and a further nine patients are described.
4. The embryology, natural history and treatment of the condition are discussed.

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**REFERENCES**


