The enigmatic role and development of the clavicle

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Abstract

The clavicle is a particularly odd bone with regard to its unique embryological development and the specific disease processes that it may encounter. The aim of this review is to describe the normal and abnormal development of the clavicle from its embryological origins until skeletal maturity. The review also attempts to account for the purpose of the clavicle by describing its structure and function.

Keywords clavicle; cleidocranial dysostosis; development; function; pseudarthrosis

Introduction

The clavicle is a bone like no other in the human body. It is the first bone to start ossification and the last one to complete the process. It develops early from two primary ossification centres and later from two secondary ossification centres. It is unknown why this ossification process is interrupted in cases of congenital pseudarthrosis, or why this condition should be four times more common in girls and occur mainly on the right side. Partial or complete absence of the clavicle is seen in cleidocranial dysostosis but, paradoxically, shoulder function is well preserved in affected individuals. In order to understand the function of the clavicle, it is important to know how the clavicle develops *in utero*, and which structures are connected. Further clues to the function of the clavicle may be gained from understanding the varying effects of congenital and acquired deformities.

Embryology

During embryological development, the clavicle forms by both intramembranous and endochondral ossification processes, which is unlike any other long bone. There are two primary ossification centres that form from mesenchymal tissue after five weeks gestation, and these rapidly fuse in the middle of the clavicle. Growth in this area occurs by intramembranous ossification. At the medial end, however, there is a secondary ossification centre. The medial physis reliably appears by the age of 17 years and fuses with the shaft between the ages of 21–24 years. It is the last bone to undergo physeal closure, which is a characteristic that makes the clavicle useful in forensic medicine to help determine age.[1]

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Eighty percent of the clavicle's length is gained by eight years of age in girls, and 12 years in boys. The right clavicle is also usually slightly larger than the left.

There are several congenital abnormalities that affect the clavicle, some of which may be inherited via a defect in the Runx-2 gene. The most notable defect is cleidocranial dysostosis, where there is absence of the clavicles.

Structure

The S-shaped clavicle has many muscular attachments including sternohyoid, sternocleidomastoid, pectoralis major, subclavius, deltoid and trapezius. Medially, the clavicle is anchored to the ribs by the costoclavicular ligament. Laterally, the conoid and trapezoid ligaments hold the clavicle down to the coracoid process of the scapula. The acromioclavicular ligaments provide anteroposterior stability to the joint and are reinforced superiorly by fibres of deltoid and trapezius. The clavicle articulates with the sternum medially, and the acromion of the scapula laterally. Although both articulations are diarthrodial joints, the sternoclavicular joint is a saddle joint whereas the acromioclavicular joint is a plane synovial joint. Both joints also contain a small fibrocartilaginous disc that acts as a passive stabilizer and connects to the surrounding ligament complexes. The clavicle is referred to as a long bone, as it is longer than it is wide and has two ends. Contrary to popular belief, it contains some bone marrow within an intramedullary canal.[2]

Function

In order for the arm to abduct 180 °, the clavicle must allow movement at both the sternoclavicular and acromioclavicular joints. This is so that the scapula can change orientation during shoulder movement and adapt to the changing contour of the chest wall. Humans are not the only vertebrates to have clavicles; in birds they are fused and often referred to as the 'wishbone', whose function is to strengthen the thorax against the harsh forces of flight. As humans are in a different taxonomic class to birds, the evolutionary advantage of clavicles must be important to vertebrates in general. It links the axial to the appendicular skeleton and has several bony functions:

- A strut that prevent the shoulder collapsing into the body
- A stabilizer to prevent the shoulder from falling away from the body
- A shield to protect the subclavian artery and brachial plexus
- A suspension for the scapula

Whether the clavicle acts as a strut or a stabilizer will depend upon the position of the ipsilateral upper limb and how the shoulder is being loaded.[3]

The medial ligament complex is made up of a large anterior costoclavicular ligament and a smaller posterior sternoclavicular ligament.[4,5]

Laterally, the clavicle attaches to the scapula through the coracoclavicular and acromioclavicular ligaments. These ligaments are important to prevent dyskinesis of the scapula.[6]

Cleidocranial dysostosis (CCD)

This condition primarily affects the development of bones and teeth[7] and has a global incidence of 1 in 1 000 000. The skull

exhibits delayed membranous ossification and the fontanelle remains open, whilst the sinuses are underdeveloped.[8] Children suffer recurrent ear infections and have retention of deciduous dentition. The clavicle has a bilateral dysplastic appearance, or sometimes is completely absent altogether. Individuals with CCD can often bring both shoulders together to touch at the midline (Figure 1). The knees develop genu valgum, the hands display brachydactyly and the spine may have a scoliosis. The *RUNX2* gene has been implicated[9,10] in cleidocranial dysostosis, it coding for a transcription factor protein that regulates osteoblasts. Although it is an autosomal dominant condition, *de novo* mutations also occur. It is still not fully understood, as the *RUNX2* gene is normal in a third of cases of CCD. Diagnosing this condition is achieved using clinical features and skeletal survey radiographs. Where there is still doubt, genetic analysis is performed. *In utero* diagnosis is possible after 13 weeks if there is delayed ossification of the vertebrae. Treatment for this condition is aimed at providing good dental care and correcting osteopaenia with calcium and vitamin D supplements.

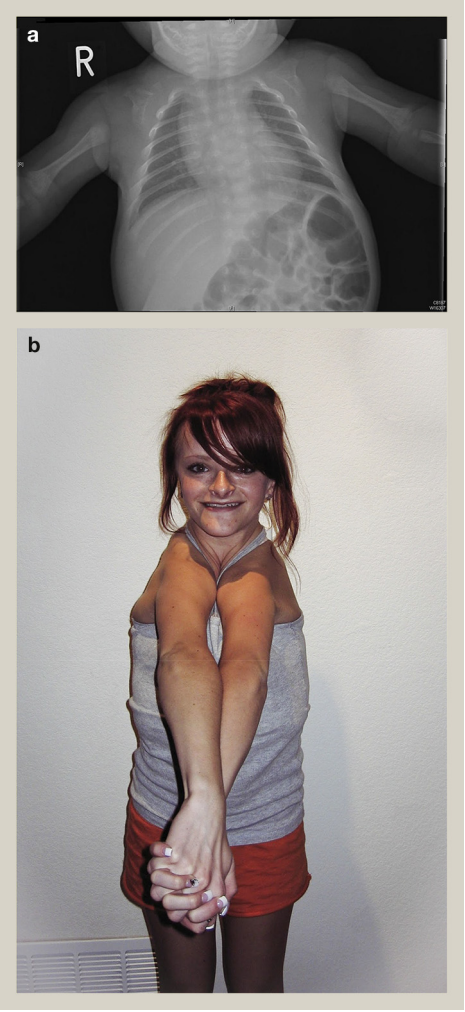

Figure 1 a: Cleidocranial dysostosis presented in a 6 week old girl with hypermobile shoulders. A chest radiograph shows hypoplasia of the clavicles. (Case courtesy of Dr Hugh Harvey, Radiopaedia.org). **b**: A clinical photograph of a young female who has CCD. She is able to touch both shoulders in the midline. (Image courtesy of Ms Whitney Madison).

Congenital pseudarthrosis of the clavicle (CPC)

This abnormality tends to occur on the right side in the majority of cases.[11] When on the left, it is associated with dextrocardia.[12] Failure of fusion of the medial and lateral primary ossification centres is thought to be the reason for developing pseudarthrosis, although the cause is still unknown. One theory suggests that pressure from the subclavian artery prevents the ossification centres from fusing, this being located higher on the right side than the left.[11] A bump is noted over the clavicle in the region of the pseudarthrosis and often becomes more prominent as the proximal end is pulled up by the sternocleidomastoid muscle. Most sufferers

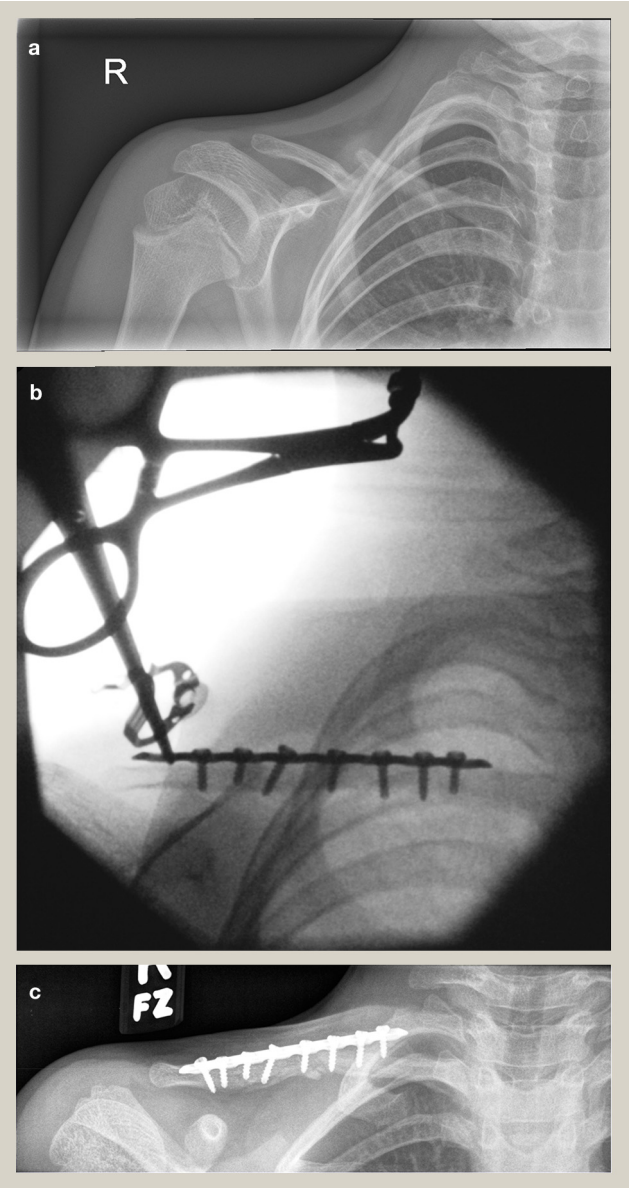

Figure 2 a: A right sided congenital pseudarthrosis of the clavicle. Both ends are smooth which indicates that this is not an acute fracture. The proximal portion of the clavicle is elevated due to the pull of sternocleidomastoid. **b**: Open reduction and internal fixation of the clavicle, using a small fragment plate and a tricortical wedge of iliac crest bone graft. **c**: The previous pseudarthrosis site has now fused, despite screw pull-out at the medial end of the plate.