Congenital Differences About the Elbow

Scott H. Kozin, MD[a,b,*]

KEYWORDS

• Congenital • Elbow • Synostosis • Arthrogryposis
• Pterygium • Multiple hereditary exostosis

Certain congenital differences immediately impair elbow motion and function, whereas other ailments slowly limit elbow motion or cause instability. Diagnoses that immediately impair elbow function are: synostosis (including proximal radioulnar, radiohumeral, ulnohumeral), arthrogryposis, and pterygium. Congenital differences that affect elbow motion or stability over time include: multiple hereditary exostosis, Ollier disease, osteogenesis imperfecta, skeletal dysplasia, nail patella syndrome, and Gorham disease. In either case, the child's ability to perform activities of daily living and to participate in recreation is diminished. The impairment is related to the deficit in elbow or forearm motion, the overall length of the limb, the degree of ipsilateral hand function, and the status of the contralateral limb. This article discusses congenital differences that affect the elbow, with an emphasis on diagnosis and treatment. In some cases, surgery is effective in enhancing function. In other cases, observation is preferable because children possess remarkable ability to adapt without surgery.

CONGENITAL DIFFERENCES THAT DIRECTLY IMPAIR ELBOW FUNCTION

Synostosis

Synostosis is a generic term that indicates an osseous union between bones that are normally separated.[1–3] Clinically, however, there is a spectrum of presentation that varies with the degree of joint development and the amount of synostosis. In other words, the motion can range from full, unrestricted movement to no motion in cases when the bones are fused. The variability of motion is most notable about the forearm joint. Many children present to the office with limited pronation or supination. Mild deficits often are unrecognized until the child is older when the necessary activities of daily living become more complicated (eg, toileting, self-feeding). A history of trauma may or may not be present. Radiographs may be normal or demonstrate subtle findings about the radiocapitellar and proximal radioulnar joints (**Fig. 1**). An understanding of the continuum from normal anatomy to rigid synostosis prevents an unnecessary work-up and unsuccessful surgery.

Synostosis across the elbow can be in a longitudinal direction (ie, ulnar–humeral or radial–humeral) or in a transverse path across the proximal radioulnar joint.[1,2] Radiohumeral synostosis most commonly occurs in ulnar deficiency, but can be part of a synostosis syndrome, such as Herrmann multiple synostosis syndrome.[1,4] Proximal radioulnar joint synostosis can occur in isolation or can be found in a variety of syndromes, including trisomy (13 or 21) and fetal alcohol syndrome.[1]

Radiohumeral synostosis

Radiohumeral or ulnohumeral synostosis impairs elbow flexion and extension. The functional impact depends upon the elbow position, hand function, compensatory surrounding joint motion, and status of contralateral limb. Because radiohumeral synostosis most commonly occurs in children with ulnar deficiency (type IV), bilateral involvement and hand impairment are prevalent.[1] The child with ulnar deficiency is often born with the hand against

[a] Department of Orthopaedic Surgery, Temple University, Philadelphia, PA, USA
[b] Upper Extremity Center of Excellence, Shriners Hospitals for Children, 3551 North Broad Street, Philadelphia, PA 19140, USA
* Shriners Hospitals for Children, 3551 North Broad Street, Philadelphia, PA 19140.
E-mail address: skozin@shrinenet.org

Hand Clin 25 (2009) 277–291
doi:10.1016/j.hcl.2008.12.007
0749-0712/08/$ – see front matter

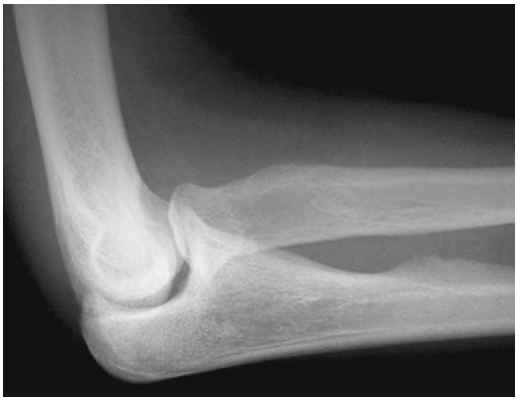

Fig. 1. A 14-year-old child presents with limited arc of forearm rotation (active pronation 45° and active supination 40°). Lateral radiograph reveals abnormal development of radiocapitellar joint with enlarged flattened radial head. (*Courtesy of* Shriners Hospital for Children, Philadelphia, PA; with permission.)

the flank, which positions the hand facing backward (**Fig. 2**). Treatment recommendations have varied from observation to corrective osteotomy to reposition the limb into a more functional location. In the past, I have improved limb position by way of osteotomy through the fusion mass. This can be accomplished acutely at the time of surgery or slowly with distraction osteogenesis. I have noticed, however, that children born with their hands facing backward often achieved similar positioning as those children who underwent surgery. In addition, their functional limitations were further lessened by their amazing compensatory motion (**Fig. 3**). The ability to compensate is unclear, but may be related to excessive surrounding joint motion or forces across the growth plate changing bony torsion. Nonetheless, the results are impressive and I now reserve osteotomy for recalcitrant cases that present with persistent poor limb position despite growth and development.

The limb with radiohumeral synostosis is often short owing to lack of growth plates. Limb lengthening has been recommended to improve workable reach space. However, distraction lengthening must be approached with great caution because the complication rate is high and the preoperative goals often unobtainable. Furthermore, limb lengthening can move the hand further from the face and adversely impact function. Therefore, considerable thought and evaluation are necessary before limb lengthening. An occupational therapy evaluation is invaluable to include an assessment of preoperative goals to prevent disappointment to both the patient and the surgeon.

Proximal radioulnar joint synostosis

Proximal radioulnar joint (PRUJ) synostosis is bilateral about 50% of the time and may be inheritable (**Fig. 4**).[2,3,5,6] Forearm motion must be assessed at the radial styloid and ulnar head (distal radioulnar joint) to avoid measuring wrist motion instead of forearm movement (**Fig. 5**). Anteroposterior and lateral elbow radiographs are necessary because the synostosis is nearly always about the PRUJ (**Fig. 6**). In the young child, the connection is initially cartilaginous but eventually ossifies to a bony connection. A concomitant radial head dislocation may be present. Advanced imaging studies are not necessary.

Shoulder and wrist motion is able to compensate for a lack of forearm rotation during many activities of early childhood. Shoulder abduction compensates for deficient pronation and shoulder adduction compensates for deficient supination (**Fig. 7**). Additionally, children with deficient forearm rotation will develop excessive wrist intercarpal rotation.[5] Therefore, a delay in presentation is common until the complexities of daily activities amplify, such as catching a ball, self-feeding, or toileting (**Fig. 8**). Common complaints from a pronation deficit are difficulties with keyboarding and tabletop activities. Common difficulties from a supination deficiency are eating, washing one's face, catching a ball, and using a soap dispenser. Many children present after unrelated trivial trauma, which alerts the parents to the absent forearm rotation.

Fig. 2. A 4-year-old child with bilateral ulnar deficiencies and right hand facing backward. (*Courtesy of* Shriners Hospital for Children, Philadelphia, PA; with permission.)