Epiphyseal Osteochondromas of the Upper Limb: A Report of 7 Cases

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Background: Originally described as osteochondromatous lesions arising from the tarsal bones, osteochondromas arising from the epiphysis or carpal/tarsal bones are less common than those arising from the metaphysis. Histologically, all osteochondromas are indistinguishable regardless of the location from which they arise. Few case reports and case series exist describing these lesions in the upper limb.

Methods: We review 7 cases of osteochondromas arising from epiphyses and ossicles in the upper limb treated at 3 institutions. Patients were followed for an average of 5.7 years. The average patient age at the presentation was 7.8 years.

Results: We identified 25 lesions: 5 distal radial epiphyseal, 3 distal radial metaphyseal, 4 scaphoid, 4 lunate, 4 trapezial, 2 accessory ossicles adjacent to the trapezium, 2 trapezoid, and 1 metacarpal lesion. Three patients presented with pain, 5 with decreased motion, and 3 with angular deformity. In 1 case, the lesion presented as an incidental finding. Four patients underwent a total of 7 procedures: 2 open biopsies, 2 distal radial epiphyseal lesion excisions, 2 revisions, and 1 excision of all lesions with a scaphoid osteotomy.

Conclusions: Intra-articular and transosseous lesions are more likely to result in angular deformities and loss of motion at the joints, whereas juxtaphyseal and transphyseal lesions are more likely to result in growth disturbances and angular deformities at the physis.

Level of Evidence: Case series, level IV.

Key Words: Trevor disease, osteochondroma, epiphysis, tarso-epiphyseal aclasis, dysplasia epiphysealis hemimelica

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Osteochondromatous lesions arising from tarsal bones were first described in 1926 by Mouchet and Belot.[1] By 1950, Trevor[2] had collected 10 cases of "tarsomegalie" and renamed the lesions "tarso-epiphyseal aclasis," recognizing that the epiphysis could also be involved. In 1956, Fairbank,[3] who had supplied several of the cases for Trevor original manuscript, reported an additional 14 cases. Until that time, all reported cases had involved 1 side of the epiphysis only; Fairbank thus proposed the descriptive term *dysplasia epiphysealis hemimelica*, which is more commonly used today. Since that time, more than 130 cases have been reported, only 25 of which have involved the upper extremity.[4–23]

The presentation and treatment algorithms for osteochondromas of the epiphysis are similar to those for osteochondromas more classically arising from the metaphysis. Indications for surgery have included pain, an enlarging mass, angular deformities, limb-length discrepancies, and carpal instability.[5,24] Recurrences are rare, and no malignant transformations have been reported.

Histologically, the lesions are indistinguishable from metaphyseal osteochondromas. They can occur as solitary lesions or multiple lesions and can coexist with solitary or multiple metaphyseal osteochondromas.[4]

The incidence is reported to be approximately 1:1,000,000, although some authors have suggested that the incidence might be much higher. No familial predisposition has been shown, and males are more commonly affected. African Americans have a reportedly lower incidence compared with whites. The lesions tend to involve only 1 extremity, even when multiple osteochondromas are present, and are more common on the radial side of the upper extremity.[25]

METHODS

Approval for a retrospective chart review of patients treated for epiphyseal osteochondromas was obtained from the institutional review board. Twenty cases of epiphyseal osteochondroma were identified at 3 institutions from 1952 to 2006, 7 of which involved the upper extremity. Complete medical records were available for all 7, including imaging studies, operative notes, and pathology reports. Patients were followed for an average of 5.7 years (range, 1 to 14 y). The average patient age at presentation was 7.8 years (range, 17 mo to 23 y). Plain radiographs were obtained of all patients during follow-up. Magnetic resonance images were obtained of 4 patients.

RESULTS

Two of the study patients had solitary lesions: 1 confined to the lunate and 1 in the trapezium. No patient

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had a family history of either epiphyseal or metaphyseal osteochondromas. A total of 25 lesions included 16 carpal osteochondromas, 6 epiphyseal osteochondromas, and 3 metaphyseal osteochondromas. The majority of the lesions involved the carpal bones: 4 scaphoid, 4 lunate, 4 trapezial, and 2 trapezoid exostoses. One patient had 2 accessory ossicles adjacent to the trapezium. Five lesions arose from the distal radial epiphysis, 3 of which were accompanied by lesions of the distal radial metaphysis. One patient had a lesion arising from the epiphysis of the first metacarpal. Three patients presented with pain. Decreased range of motion of the wrist was noted in 5 patients, with angular deformity of the wrist and pain with an enlarging mass present in 3 of the 5 patients. Another patient presented with pain and a mass of increasing size at the wrist without deformity.

All patients had plain radiographs available for review, and 4 patients had also undergone magnetic resonance imaging (MRI) of the involved area. Radiographic findings included focal overgrowth of the distal radial epiphysis and/or the carpal bones and pedunculated lesions consistent with osteochondromas (Fig. 1). MRI revealed that many of the lesions were contiguous, spanning the physis or the intercarpal joints. In one patient, thin excrescences were observed bridging the exostoses of the distal radius and those of the trapezium and capitate. Another patient had two 1- to 2-mm-wide ossific bridges connecting the metaphyseal and epiphyseal exostoses, with cartilage covering the underlying irregularly contoured physis (Fig. 2).

Seven surgical procedures were performed on 4 of the 7 patients. Two patients underwent open biopsy: 1 for an enlarging mass with pain and 1 for an atypical mass spanning the distal radial physis involving both the metaphysis and epiphysis. Two patients underwent excision of the distal radial epiphyseal lesions: 1 for an ulnar deviation deformity and the other for decreased wrist extension. Of those, the patient with the angular deformity experienced worsening of the condition and required 2 subsequent operations for excision of the carpal lesions. The other patient achieved improvements in motion, but pain had recurred. One patient underwent excision of all lesions simultaneously along with a wedge osteotomy of the scaphoid at an older age (9 y) and remained pain-free and unlimited in activities despite decreased wrist motion. No patient underwent lengthening or shortening for limb-length discrepancy.

At final follow-up, all patients who initially had presented with pain continued to have pain with activities. By contrast, only 1 patient who presented without pain complained of pain at final follow-up. At the time of this study, that patient was awaiting a realignment procedure for ulnoradial length discrepancy. One patient who had presented with a painless mass at the age of 5 years did develop pain during follow-up and eventually underwent excision of all the osteochondromas in a single procedure at the age of 9 years. He was pain-free at the last follow-up visit. When present, pain was managed with intermittent splinting, rest, ice, and non-narcotic pain medications.

DISCUSSION

Osteochondromas of the epiphysis or carpal bones are rare, and such osteochondromas of the upper extremity are even rarer. To date, only 25 cases involving the upper limb have been reported.[4–23] Most cases of epiphyseal osteochondromas arise from the distal radius, and most also involve the carpal bones. All 7 of our

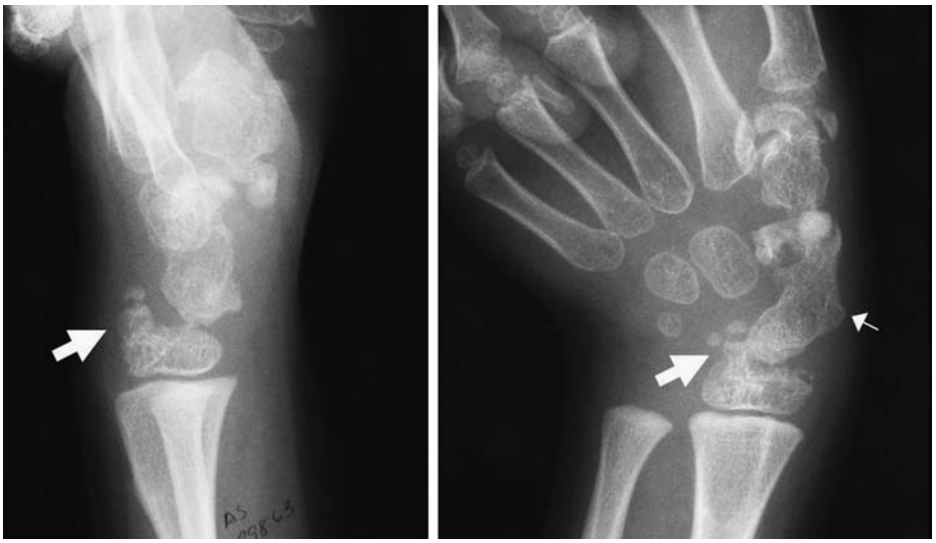

FIGURE 1. Anteroposterior and lateral radiographs of a child's wrist demonstrating multiple enchondromatous lesions, including pedunculated (thick arrow) and sessile (small arrow) forms.

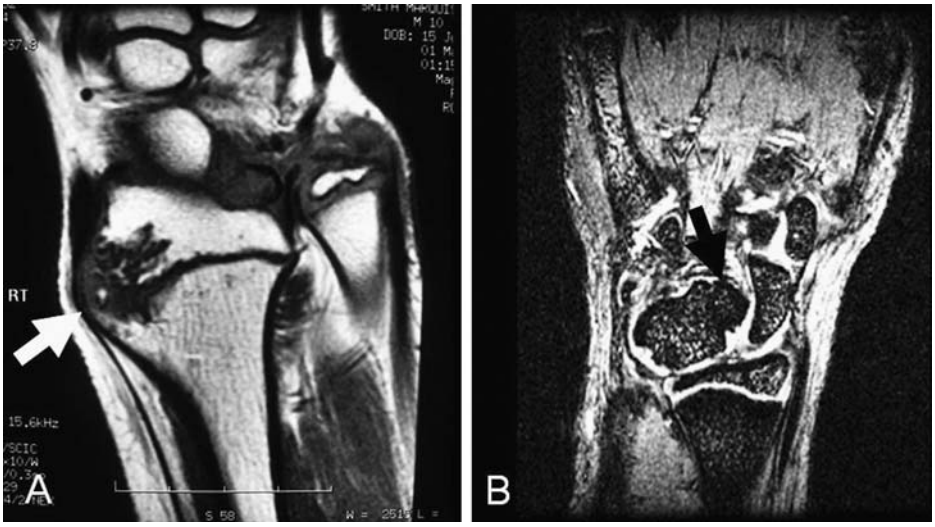

FIGURE 2. A, Coronal T1-weighted MRI demonstrating a transphyseal lesion spanning the distal radial epiphysis. Note the expanded and irregularly contoured physis adjacent to the apparent physeal bridge (white arrow). B, Coronal T2-weighted MRI image demonstrating a large solitary osteochondroma of the lunate (black arrow). The more radial carpals are spared. MRI indicates magnetic resonance imaging.

patients had carpal involvement, and all epiphyseal osteochondromas except 1 were localized to the radial side of the wrist. We found no predilection for any ethnic background in our small series. The male to female ratio was nearly 2:1.

Only 2 of our patients had solitary lesions, and all 3 of the patients with multiple epiphyseal osteochondromas who underwent MRI were revealed to have interconnections between the lesions, either transphyseal or transosseous. Interconnected lesions in the carpus have previously been described[5] and might be responsible for the decreased intercarpal and radiocarpal distance shown on radiographs (rather than an advance in bone age). Bony or cartilaginous bridging lesions might be evident only on magnetic resonance images; therefore, the incidence might be underreported.

Imaging studies include plain radiography. MRI might be indicated to define the intra-articular lesions. Asymptomatic lesions need only be observed, considering that no malignant transformation of epiphyseal osteochondromas has been reported.

Patients whose primary complaint is pain, with or without mechanical symptoms, should be counseled that they are likely to have persistent pain despite operative intervention. Those patients with angular deformities, mechanical blocks to motion, or growth disturbances might benefit from osteotomies, resection of the mass, or bone lengthening or shortening, respectively. None of our patients had growth disturbances that required surgical intervention. Patients with bony blocks to motion did improve after excision or debulking of the lesions, although 1 patient required 2 reoperations for recurrence of deformity.

On the basis of this small series of cases, it seems that treatment was more effective if delayed until pain or loss of motion interfered with function. Debridement of the osteochondromas can be combined with corrective osteotomy if deformity correction is needed. The child whose intra-articular bony excrescences underwent debridement in combination with a closing wedge osteotomy improved both position and motion. Removal of a lesion that bridges the physis did not interfere with growth in one child but did not prevent additional growth retardation in another.

Epiphyseal osteochondromas are histologically identical to metaphyseal osteochondromas and might represent different modes of expression of the same pathologic process.[4,26] The variable might be the location from which the cartilaginous proliferation arises and might represent a continuum from the metaphysis to the physis and around the entire epiphysis. Metaphyseal lesions have the appearance of growing away from the physis as normal physeal growth leads to bone elongation. Epiphyseal lesions can radiate from anywhere along the epiphysis and might therefore extend into the joint, grow alongside the joint, emerge from the physeal margin, grow across the physis, or connect to other bones in the region.

Rather than Trevor disease, we have used the term *epiphyseal osteochondroma*, either solitary or multiple, as suggested by Goldenberg in 1966.[24] This name, however, does not adequately describe the lesions that involve the small bones of the carpus or tarsus in that those do not have epiphyses in the true sense. However, carpal and tarsal bones grow in a manner similar to that of epiphyses, which might explain why the lesions occur

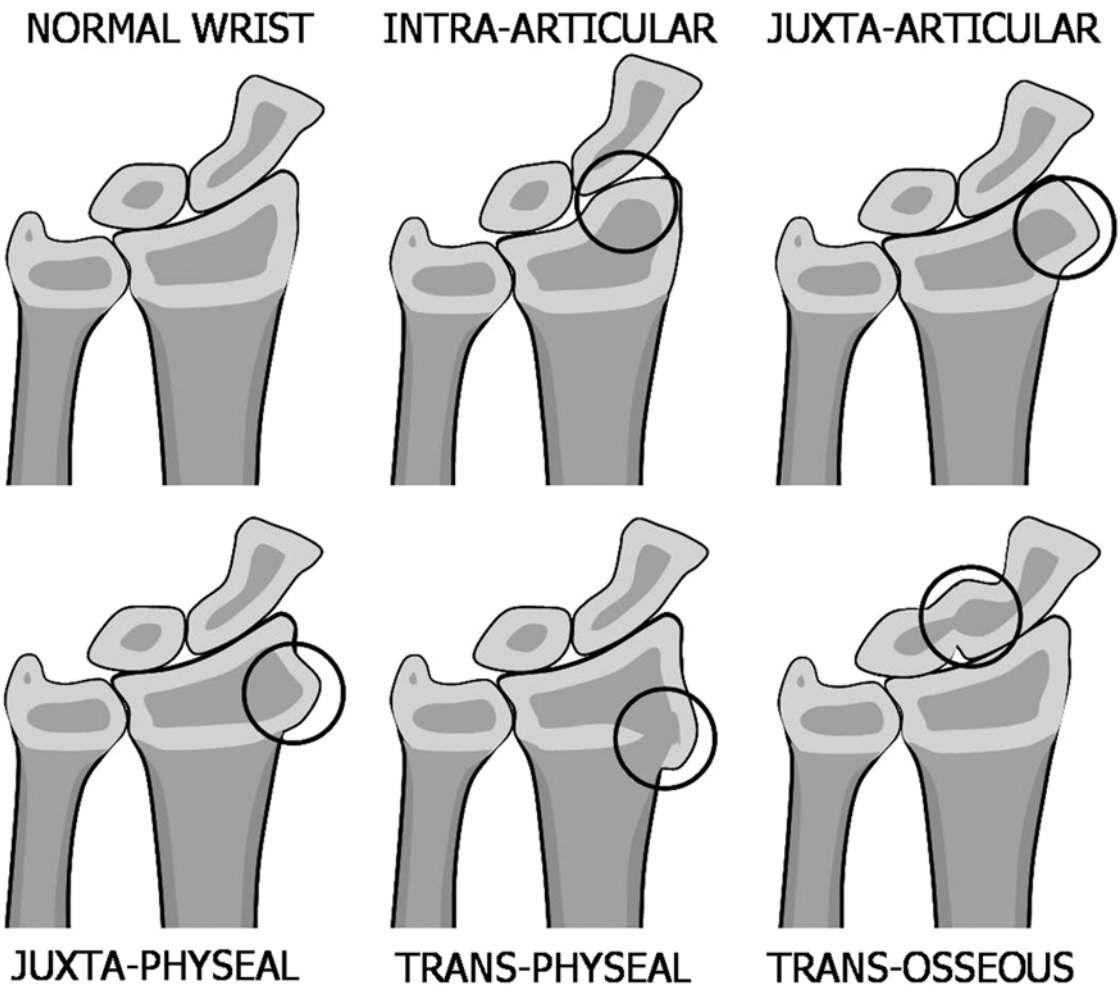

FIGURE 3. Diagrammatic representation of the various types of osteochondromas encountered at the wrist. These same types can also be used to subdivide epiphyseal osteochondromas in other anatomic regions. Intra-articular and transosseous lesions manifest clinically with loss of motion and/or pain, whereas juxta-articular lesions may have a benign course. Transphyseal lesions behave analogously to physeal bars, presenting with growth disturbances or angulation. Juxtaphyseal lesions may or may not result in growth disturbances, and may represent a precursor to transphyseal lesions.

together. A more accurate way of describing these lesions might be to group all osteochondromas together and then subdivide them according to location relative to the ossification center of the epiphysis or the carpal or tarsal bone.

Kuo et al[25] proposed classifying the lesions as either juxta-articular or intra-articular. On the basis of our MRI findings, we propose 4 further subgroups: metaphyseal, transphyseal, juxtaphyseal, and transosseous lesions (Fig. 3). We therefore suggest a comprehensive classification scheme of all osteochondromas to incorporate our findings and those of previously reported studies (Table 1).

The most common lesion involving the carpal bones and epiphyses both in our series and in the review of the literature was the intra-articular type, affecting 26 of 30 patients with adequate radiographs for evaluation. Some selection bias might exist, however, in that patients with extra-articular lesions might be more likely to be asymptomatic and not seek medical attention. One of our cases with a juxta-articular lesion presented as an incidental finding.

Using our comprehensive classification to characterize our results and those of previously reported cases, intra-articular and transosseous lesions are more likely to result in angular deformities and loss of motion at the joints, and juxtaphyseal and transphyseal lesions are more likely to result in growth disturbances and angular deformities at the physis. Juxta-articular lesions are the most benign and generally result in a painless mass or only local irritation of traversing soft-tissue structures.

TABLE 1. Universal Classification of Osteochondromas

Classification	Typical Presentation	Imaging Studies	Treatment
I. Metaphyseal	Painless lump or local soft tissue irritation	XR: Pedunculated or sessile mass arising from the metaphysis growing away from the physis MRI: Intramedullary canal of lesion continuous with canal of adjacent bone	Excision if causing pain or limiting motion
II. Transphyseal	Growth arrest and/or angular deformity	XR: May resemble physeal bar but with associated bony mass along physis MRI: Expanded and irregularly contoured physis adjacent to apparent physeal bridge	Excision if limiting growth or causing angular deformity (similar to peripheral physeal bar resection), may require correctional osteotomy depending on age
III. Juxtaphyseal	Painless lump or local soft tissue irritation, possible angular deformity	XR: Pedunculated or sessile mass arising from the epiphysis growing away from the physis MRI: Intramedullary canal of lesion continuous with canal of bone from which it arises	Excision if causing pain or limiting motion, may require correctional osteotomy depending on age
IV. Juxta-articular	Loss of joint motion and/or angular deformity	XR: Lesion may abut or displace carpal bones MRI: In younger children with unossified carpal bones, mass effect better visualized on MRI	Excision if causing pain or limiting motion
V. Intra-articular	Loss of joint motion and/or angular deformity	XR: Mass effect will displace adjacent carpal bones MRI: In younger children with unossified carpal bones, mass effect better visualized on MRI	Excision if causing pain or limiting motion, may require limited intercarpal fusions/osteotomies to address deformity and loss of articular cartilage of carpal bones
VI. Transosseous	Loss of joint motion and/or angular deformity	XR: May resemble carpal coalition but with associated bony mass and irregular contours MRI: Expanded and irregularly contoured carpal bones or distal radius joined together with continuous intramedullary space	Excision if causing pain or limiting motion, may require limited intercarpal fusions/osteotomies to address deformity and loss of articular cartilage of carpal bones

MRI indicates magnetic resonance imaging; XR, radiography.

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