More Than Epiphyseal Osteochondromas: Updated Understanding of Imaging Findings in Dysplasia Epiphysealis Hemimelica (Trevor Disease)

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OBJECTIVE. The purpose of this article is to discuss approaches to imaging dysplasia epiphysealis hemimelica in the context of recent advances in the understanding of the underlying pathophysiologic profile of this entity, which may result in pain, growth disturbance, and early development of osteoarthritis.

CONCLUSION. Dysplasia epiphysealis hemimelica was first characterized as a skeletal disorder with osteochondromas characteristically involving epiphyses on one side of the same lower extremity. Upper extremity involvement was subsequently recognized. Previously conceptualized as epiphyseal osteochondromatosis, recent investigations have uncovered differences between these osteocartilaginous lesions and osteochondromas.

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Characterized by epiphyseal overgrowth and osteocartilaginous epiphyseal lesions caused by idiopathic benign cell proliferation, dysplasia epiphysealis hemimelica (DEH), also known as Trevor disease, is an infrequently reported clinical entity that may manifest with painless deformity, limited range of motion, mechanical pain, and localized growth disturbances. Imaging is essential in establishing the diagnosis and guiding the management of DEH. This article discusses approaches to imaging DEH in the context of recent advances in the understanding of the underlying pathophysiologic profile of this entity, which differs significantly from that of osteochondromas and which appears to entail greater underlying growth disturbances than has previously been acknowledged.

History

DEH has undergone multiple permutations of nomenclature, classification, and conceptualization that are worth briefly mentioning to avoid further confusion [1]. The first documented entity resembling DEH was reported in 1926 by Mouchet and Belot, who described an osteochondroma and overgrowth of the talus, an epiphyseal equivalent, and named this condition *tarsomégalie* [2]. Later, in the 1950s, Trevor [2] characterized additional cases involving the lower extremities and called the condition "tarso-epiphysial aclasis," despite the lack of tarsal involve-

ment in all cases. Fairbank [3] refined the understanding of this condition by collecting further examples and conceptualizing this entity as tending to affect the epiphyses of the same side of one lower extremity, with the condition referred to as "dysplasia epiphysialis hemimelica."

Epidemiologic Profile and Clinical Presentation

DEH is considered rare, with an estimated frequency of one case among 1 million individuals and approximately 150 reported cases in the literature; however, the accuracy of this epidemiologic estimate has been criticized because the estimated frequency based on the reported literature would be substantially higher than the reported frequency [1, 4, 5]. It is also likely that many of these cases are misdiagnosed as osteochondromas or other disease entities or are asymptomatic, making DEH more common than has been reported in the past. Male patients are predominantly affected and have an approximately threefold greater representation than female patients in the published literature [1, 6]. In most children, DEH is diagnosed during early childhood, generally before 8 years of age, with the earliest diagnosis reported for a patient aged 8 months and with presentation during adulthood considered unusual [1, 3, 7, 8]. DEH generally entails unilateral involvement of an epiphysis or epiphyseal equivalent (e.g., calcaneus, talus, patella, ischium, pubic bone, and car-
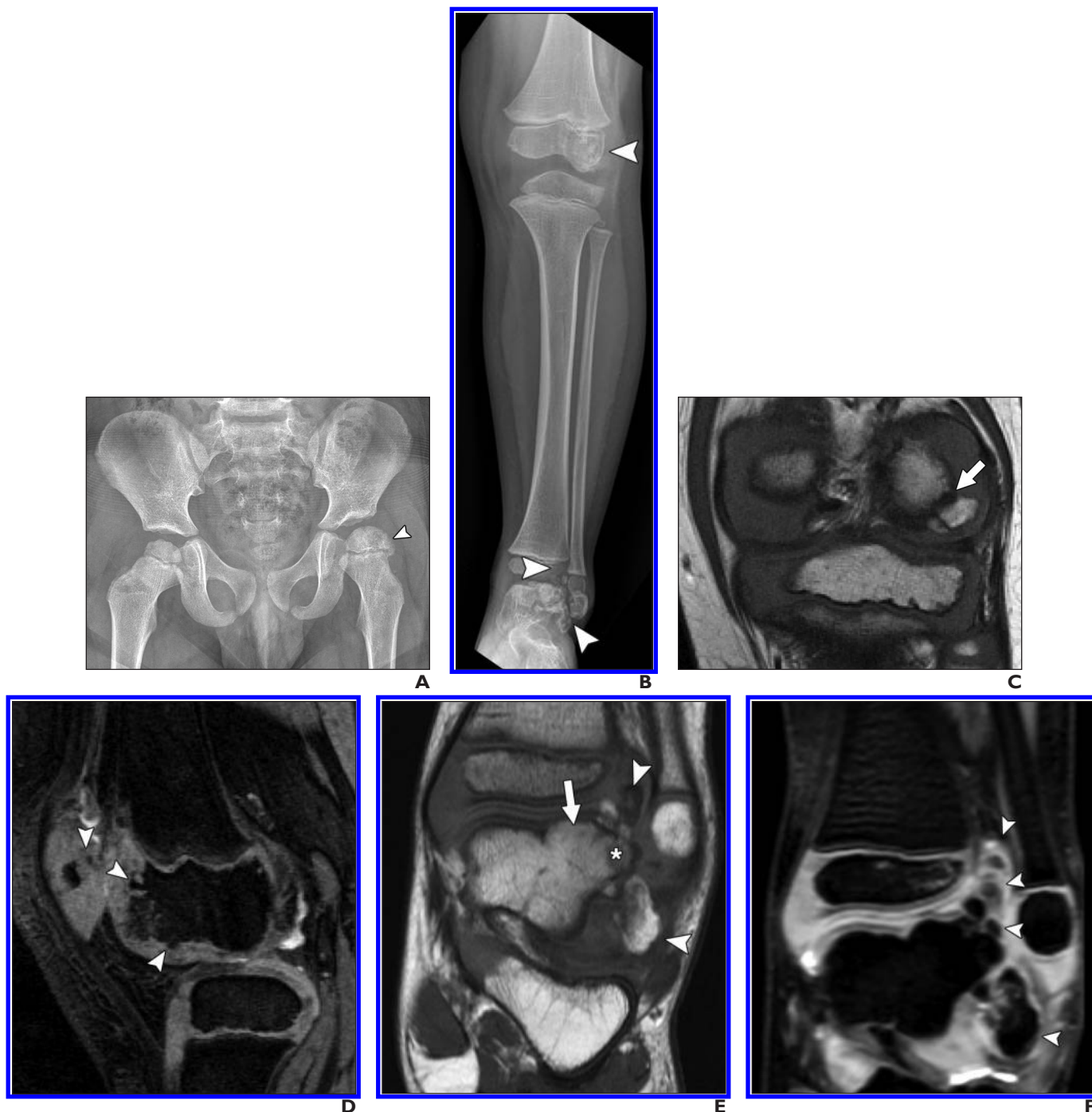
Fig. 1—1-year-old boy who presented with limp.

A, Anteroposterior (AP) radiograph of pelvis shows asymmetric enlargement and irregular ossification center that particularly involves lateral aspect of left proximal femoral capital epiphysis (*arrowhead*).

B, AP radiograph of left lower extremity also shows irregular ossific densities that involve lateral femoral condyle, distal tibia, and talus (*arrowheads*) in pattern consistent with generalized-type dysplasia epiphysealis hemimelica (DEH).

C, Coronal T1-weighted MR image shows ossification of lateral femoral condyle osteocartilaginous lesion with high T1-weighted signal. In addition, MR image identifies clear cleft between osteocartilaginous DEH lesion and underlying epiphysis (*arrow*).

D, High-resolution double-echo steady-state sagittal MR image shows extensive abnormal ossification centers that involve lateral femoral condyle, lateral tibial epiphysis, and patella (*arrowheads*) but were not appreciable on previously obtained radiographs. In addition, diffuse articular chondral thinning was observed on MRI.

E, Coronal T1-weighted MR image of left ankle shows similar abnormal ossified osteocartilaginous lesions (*arrowheads*) along with enlargement of lateral aspect of talus (*arrow*). Abnormal signal also exists within underlying body of talus with heterogeneous low signal, particularly within subchondral regions (*asterisk*).

F, Coronal double-echo steady-state MR image of ankle highlights secondary osteocartilaginous lesions (*arrowheads*) with surrounding cartilage. Pathologic analysis after talar lesion resection confirmed clinical diagnosis of DEH.

TABLE 1: Anatomic Distribution of Dysplasia Epiphysealis Hemimelica

Anatomic Region	In Rosero et al. [21]a		In Arealis et al. [1]b	
	Affected Bone	Frequency (%)	Affected Joint	Frequency (%)
Lower extremity	Distal tibia or fibula	22	Ankle	33
	Talus or calcaneus	22		
	Distal femur	21	Knee	25
	Proximal tibia	11		
	Tarsal bones	10	Foot	8
	Proximal femur	5	Hip	7
	Acetabulum	3		
Upper extremity	Scaphoid	2	Wrist	13
			Hand	5
	Scapula	1	Shoulder	3
			Elbow	3
Other			Sacroiliac	< 1
			Spine	< 1

Note—Data from [1, 21].
aPublished in 2006.
bPublished in 2014.

pal bones) or multiple epiphyses within one extremity [1]. Lower extremity involvement occurs three times more frequently than upper extremity involvement [1]. Classically, involvement may occur in a hemimelic distribution involving multiple joints on the same side of the extremity (Fig. 1), with the medial side more often involved [9]. Bilateral distributions are infrequent but have been reported [1, 10, 11]. The relative frequencies of both upper and lower extremity presentations are reported by site (Table 1). Symptoms are largely dependent on the size and location of the epiphyseal lesion, with most patients reporting combinations of deformity, swelling, restricted motion, or mechanical pain. As an example, large talus lesions may present with ankle impingement symptoms, whereas knee lesions may cause genu varus or valgus deformities (Fig. 2), depending on the side affected [1, 12].

Pathophysiologic Profile
Normal bone growth occurs in an orderly fashion, with a focal collection of hyaline cartilage noted at the articular aspect of each bone [13]. This cartilage comprises articular, epiphyseal, and physeal cartilage components. Epiphyseal cartilage normally is formed by well-organized chondrocytes that undergo hypertrophy and then ossify to form one or multiple secondary ossification centers [13]. Ossification centers initially are rounded in shape and then show flattened growth and closer apposition to the underlying metaphysis [13]. Imaging properties for unossified cartilage reflect its high water content (e.g., high fluid-sensitive T2-weighted and intermediate T1-weighted signal), whereas the secondary ossification center more closely matches bone as it matures and water content decreases (e.g., low
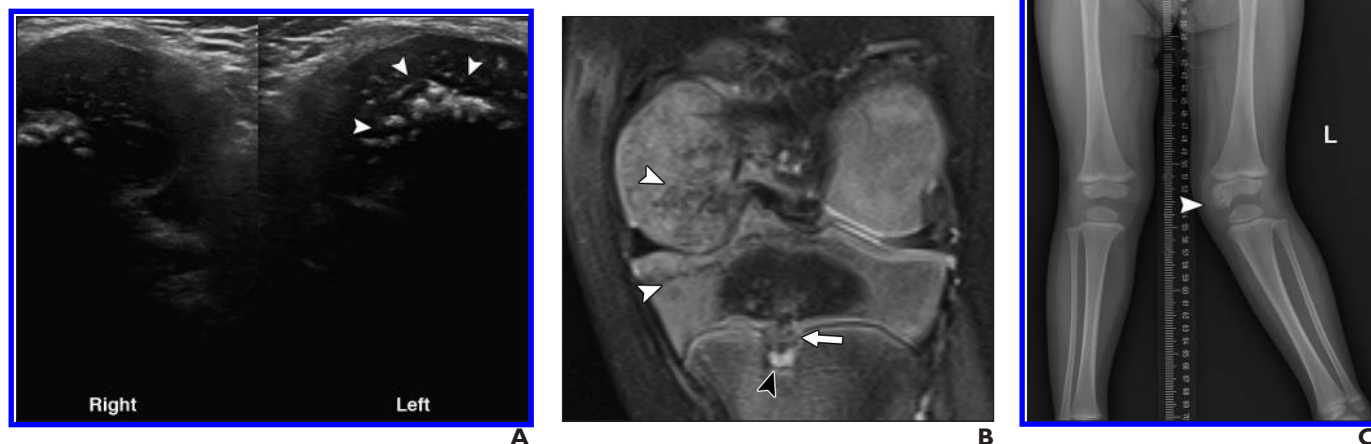

Fig. 2—8-month-old girl with left lower extremity genu valgum. Bone-length study (not shown) obtained when patient was 16 months old showed left genu valgum and abnormal ossification centers within left medial femoral condyle.
A, Transverse medial knee ultrasound images obtained soon after initial evaluation performed when patient was 8 months old shows asymmetric enlargement of left medial femoral condyle and more echogenic foci corresponding to overgrowth and additional ossification centers (*arrowheads*).
B, Follow-up coronal intermediate-weighted turbo spin-echo fat-saturated MR sequence obtained when patient was 20 months old shows overgrowth and osteocartilaginous lesions of medial knee femoral condyle and proximal tibia (*white arrowheads*). Physeal bar formation (*arrow*) developed after prior examination and was accompanied by abnormal signal within physis that was thought to represent concomitant involvement of physeal cartilage (*black arrowhead*). Findings were consistent with dysplasia epiphysealis hemimelica with physeal involvement.
C, Follow-up bone-length study (orthoroentgenogram) obtained when patient was 29 months old shows increased left genu valgum deformity, progressive ossification of medial femoral condyle lesion (*arrowhead*), overgrowth of medial femoral condyle, and proximal tibia. Limb length discrepancy (left limb length longer than right limb length) increased from 0.6 cm to 1.7 cm. L = left.

TABLE 2: Classifications of Dysplasia Epiphysealis Hemimelica

Azouz Classification From 1985 [6]	Arealis Classification From 2014 [1]
Localized: involving a single bone	1. Lower limb involving single joint
Classic: involving more than one bone of a single limb	2. Lower limb involving multiple joints
Mouchet and Belot (subtype of classic type): involving talus and medial malleolus	
Generalized or severe: involving entire lower limb from pelvis to foot	
	3. Upper limb involving single joint
	4. Upper limb involving multiple joints
	5. Upper limb and lower limb involvement
	6. Spine involvement

Note—Data from [1, 6].

fluid-sensitive T2-weighted signal), although it may contain persistent hematopoietic marrow in normal development [13].

In DEH, primarily abnormal development of the epiphyseal cartilage results in abnormal hypertrophy of the epiphyseal cartilage and subsequent excessive ossification. DEH is thought to constitute an idiopathic, sporadic epiphyseal overgrowth abnormality without any identifiable hereditary or environmental risk factors [1, 14]. Although these lesions have been frequently reported as epiphyseal osteochondromas, more recent pathologic investigations have revealed that DEH lesions are distinct from osteochondromas [15, 16]. Previously thought of as osteochondromas, these DEH lesions are more accurately described as osteocartilaginous

nodules with cartilage bands separating the nodules made up of cancellous bone [16]. In DEH, chondrocyte clusters are seen in conjunction with disorganized cartilage, unabsorbed cartilage fragments, and small ossification centers, whereas osteochondromas follow a more orderly, contiguous ossification process that more closely simulates the normal development of the physis [15]. This observation supports a newer theory that DEH represents a defect in maintaining quiescence of resident chondroprogenitor cells within the epiphysis [15, 17]. Other work also emphasizes that the gene pathways (*EXT1* and *EXT2*) implicated in osteochondromas are not detected in DEH [18]. Despite these recent advances in distinguishing DEH from osteochondromas, even recent radiologic lit-

erature continues to describe DEH lesions as epiphyseal osteochondromas.

Of importance, DEH appears to represent a primary abnormality of cartilage development that predominantly involves overgrowth of the epiphyseal cartilage, which subsequently undergoes excessive endochondral ossification during skeletal development. Given the imaging findings from more recent MRI investigations showing involvement of other nearby epiphyses as well as associated metaphyseal and physeal abnormalities, it is likely that this entity is better envisioned as a contiguous spectrum of abnormal epiphyseal cartilage development that may secondarily affect or directly involve adjacent articular cartilage, physeal cartilage, and metaphysis. The understanding of this uncommon entity is currently undergoing a revision in conceptualization, but more investigation of the radiologic-pathologic correlation is needed.

Classification

Controversies exist regarding the description and classification of DEH, most of which are attributable to the heterogeneous distribution and presentation of these lesions. Azouz et al. [6] first classified different forms of DEH into three types (Table 2): localized DEH, which affects a single bone; classic DEH, which involves more than one bone of a single lower extremity in a hemimelic distribution (on the same side of the extremity [i.e., medial versus lateral]); and generalized or severe DEH, which involves an entire lower limb from pelvis to foot. They also
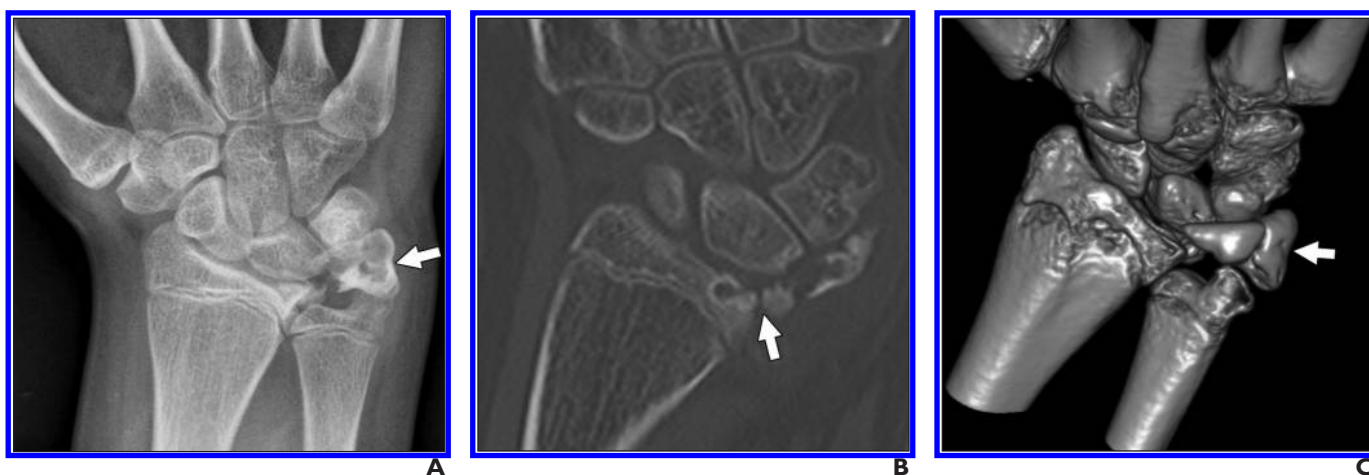
Fig. 3—11-year-old girl with wrist pain.
A, Posteroanterior radiograph of right wrist shows large lobulated ossific density involving medial aspect of wrist joint with unclear site of origin (*arrow*).
B, Coronal CT scan shows ossified osteocartilaginous lesion with fragmented appearance that appears to arise from medial aspect of radial epiphysis with clear cartilaginous cleft identified (*arrow*).
C, Three-dimensional volume-rendered reconstructed CT scan characterizes anatomic relationship of this large ossified lesion (*arrow*) before definitive resection, which confirmed dysplasia epiphysealis hemimelica.
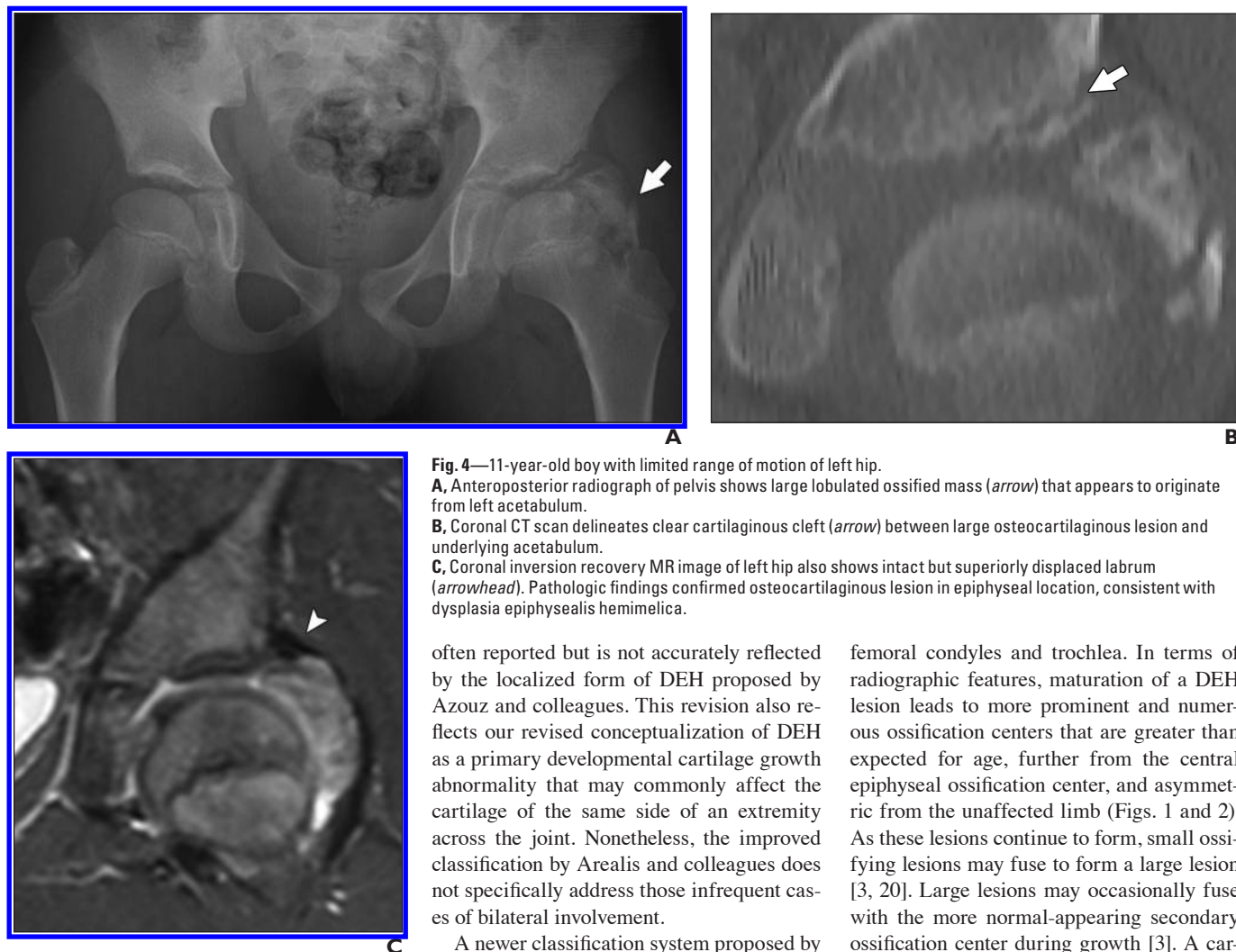
Fig. 4—11-year-old boy with limited range of motion of left hip.
A, Anteroposterior radiograph of pelvis shows large lobulated ossified mass (*arrow*) that appears to originate from left acetabulum.
B, Coronal CT scan delineates clear cartilaginous cleft (*arrow*) between large osteocartilaginous lesion and underlying acetabulum.
C, Coronal inversion recovery MR image of left hip also shows intact but superiorly displaced labrum (*arrowhead*). Pathologic findings confirmed osteocartilaginous lesion in epiphyseal location, consistent with dysplasia epiphysealis hemimelica.

created a subclassification of the classic form of DEH, known as the Mouchet and Belot type, wherein the talus and medial malleolus are involved [6].

More recently, Arealis et al. [1] noted that pitfalls in the existing classification system created by Azouz et al. [6] that result from the lack of categories for upper extremity cases or combined upper and lower extremity involvement. Although upper limb involvement is less common (Fig. 3), it is thought to occur in approximately one-quarter of cases, whereas simultaneous upper and lower limb involvement is much less common, with, to our knowledge, only three cases having been reported [1]. Therefore, Arealis and colleagues proposed their own classification, which is summarized in Table 2. The Arealis classification reports involvement on the basis of joints rather than bones, which has significant benefits because involvement across a joint is

often reported but is not accurately reflected by the localized form of DEH proposed by Azouz and colleagues. This revision also reflects our revised conceptualization of DEH as a primary developmental cartilage growth abnormality that may commonly affect the cartilage of the same side of an extremity across the joint. Nonetheless, the improved classification by Arealis and colleagues does not specifically address those infrequent cases of bilateral involvement.

A newer classification system proposed by Clarke [19] primarily distinguishes types on the basis of articular involvement and additionally incorporates bilateral involvement. This nomenclature reflects a synthesis of the traditional Azouz classification and the Arealis classification [19]. Of note, these classification systems do not differentiate on the basis of accompanying involvement of the physis or metaphysis, which may be encountered in some cases as well.

Imaging Features
Conventional Radiography

DEH traditionally shows a pathognomonic appearance on radiographs, with lobulated ossific masses, asymmetric epiphyseal enlargement, irregular ossification centers within one side (i.e., medial versus lateral) of an affected epiphysis, or a combination of these features (Figs. 1–4). In normal development, multiple ossification centers may be observed, most conspicuously within the

femoral condyles and trochlea. In terms of radiographic features, maturation of a DEH lesion leads to more prominent and numerous ossification centers that are greater than expected for age, further from the central epiphyseal ossification center, and asymmetric from the unaffected limb (Figs. 1 and 2). As these lesions continue to form, small ossifying lesions may fuse to form a large lesion [3, 20]. Large lesions may occasionally fuse with the more normal-appearing secondary ossification center during growth [3]. A cartilaginous component representing the hypertrophied, disorganized cartilage may also be appreciated on radiographic images obtained before ossification as an abnormal soft-tissue density. Extremity-only skeletal surveys and limb length studies (Fig. 2) can be helpful in screening for additional involved epiphyses and quantifying limb length discrepancy, respectively [4–6, 21]. Because these lesions may be initially asymptomatic, and because simultaneous upper and lower limb involvement have been reported, extremity-only skeletal surveys may identify additional lesions, provide comparison images for asymmetry of normal multiple ossification centers, and assist in exclusion of mimicking causes.

CT

CT provides excellent anatomic detail and can clarify the anatomic location of ossified DEH lesions, especially when they are irregular or large. DEH lesions often show a

calcified chondroid appearance that is well visualized on CT [11]. This modality may often assist in ascertaining separation from the underlying normal epiphysis [22] (Figs. 3 and 4). Three-dimensional reconstructions using volume rendering are often helpful to the orthopedic surgeon in planning for resection, to identify a cleft between the DEH lesion and an underlying more normal-appearing secondary ossification center or physis [23] (Fig. 3). However, CT is limited by its insensitivity to unossified cartilaginous components and may have difficulty delineating the relationship between abnormal osteocartilaginous lesions with the parent bone through the epiphysis, which are better appreciated on MRI [24].

MRI

MRI is particularly helpful in assessing the full extent of the cartilaginous components of the epiphyseal lesions seen in DEH [1]. Of importance, MRI assesses epiphyseal overgrowth before the ossification required

for radiographic visualization [25] (Fig. 2). Because DEH begins as a process of disorganized cartilaginous hypertrophy, asymmetric growth of the epiphyseal cartilage may be the only abnormality seen on MRI in the early stages of DEH, and this finding would not be as easily assessed on radiography or CT.

Signal characteristics of DEH lesions generally follow those of cartilage initially and then those of bone after endochondral ossification. Cartilaginous components show low-to-intermediate T1-weighted signal and hyperintense T2-weighted signal [26]. T2-weighted signal may be slightly hyperintense relative to normal cartilage, possibly reflecting disorganization [20, 25]. As ossification of the osteocartilaginous lesion progresses, areas within the epiphyseal lesion show low signal on both T1-weighted and T2-weighted sequences [25] (Fig. 1). Administration of gadolinium-based contrast medium is generally unrevealing without observation of significant enhancement, although some authors report its utility when lesions are potentially complicated by infection or inflammation [27]. Because these lesions can sometimes raise clinical concern for a malignancy or because the interpreting radiologist is unfamiliar with this entity, gadolinium administration is often performed, but no clear diagnostic benefit exists.

MRI also plays a critical role in establishing the status of the physis because premature physeal fusion (Fig. 2) or remodeling of the physis has been reported with DEH and can

also characterize the metaphyseal overgrowth or irregularity seen in some cases [6, 26] (Fig. 5). Establishing physeal involvement is important in planning for surgical intervention. Specifically, identification of cartilaginous clefts between the osteocartilaginous lesion and an underlying relatively normal-appearing secondary ossification may be critical for allowing resection while promoting continued limb growth in these skeletally immature children. Metaphyseal involvement in DEH has been mistakenly conceptualized as either concomitant development of a typical metaphyseal osteochondroma or metaphyseal enchondroma formation, such as in Ollier disease [28, 29]. However, given the contiguity of these processes, it is more likely that metaphyseal involvement reflects an extension of the same disease rather than two concomitantly occurring separate uncommon entities. The more recent recognition of DEH as involving cartilaginous disorganization and hypertrophy before endochondral ossification (dissimilar from osteochondromas) may explain the metaphyseal, physeal, and cartilaginous abnormalities observed in DEH that have been recognized on MRI only more recently. We posit that the hypertrophic, disorganized cartilage encountered in DEH occurs along a spectrum wherein milder cases involve epiphyseal cartilage only but more severe cases also affect the adjacent physeal cartilage or even extend into the metaphysis.

Articular cartilage injury or thinning (Fig. 1) that is related to large DEH osteocartilagi-

Fig. 5—11-year-old boy with chronic ankle pain and swelling.
A, Mortise view radiograph of right ankle shows irregularity of lateral aspect of distal tibia, including abnormal striated and asymmetrically enlarged appearance of lateral metaphysis (*arrow*). Premature partial physeal fusion and overgrowth is seen, including distal tibial metaphysis. There is also overgrowth of talus.
B, Coronal STIR MR image shows abnormal fusion of distal tibial physis laterally (*arrowhead*) with overgrowth of epiphysis extending into metaphysis. This abnormal growth appears contiguous from epiphysis through metaphysis laterally.
C, Sagittal T2-weighted fat-saturated MR image shows increased bursal fluid posteriorly (*arrow*) in setting of large dysplasia epiphysealis hemimelica osteocartilaginous lesion. Pathologic analysis after resection of talar lesion confirmed diagnosis of dysplasia epiphysealis hemimelica. Epiphyseal and metaphyseal involvement shown in this case is favored to represent contiguous involvement of abnormal cartilage development rather than coincidental simultaneous metaphyseal enchondroma formation as previously suggested by others.
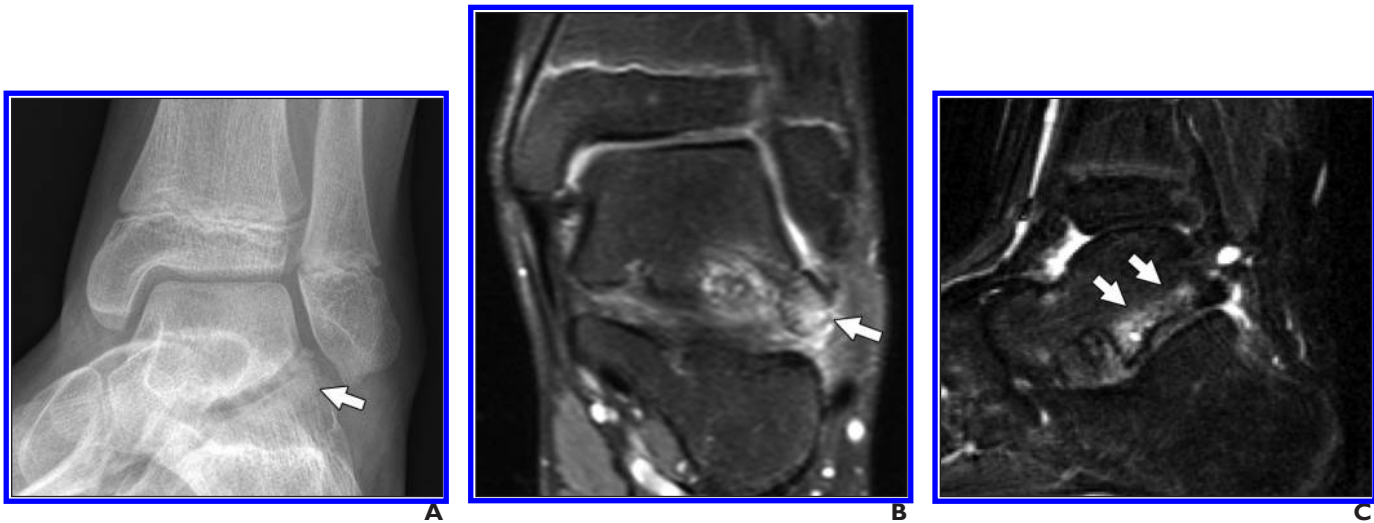
Fig. 6—10-year-old girl with recurrent ankle pain.
A, Oblique radiograph shows abnormal ossific densities near lateral aspect of talus (*arrow*).
B, Coronal proton-density imaging shows hyperintense signal involving pedunculated ossified lesion arising from lateral talus (*arrow*).
C, Sagittal STIR MR image also shows marrow edema involving posterior talus (*arrows*), likely secondary to impingement related to large dysplasia epiphysealis hemimelica lesion. Pathologic analysis confirmed osteocartilaginous lesion in epiphyseal location, consistent with dysplasia epiphysealis hemimelica.

nous lesions is also detectable on MRI, some instances of which may reflect local biomechanically mediated injury resulting from increased stress on these articulations caused by underlying epiphyseal enlargement resulting from asymmetric pressure on the affected side [26].

MRI additionally evaluates nearby osseous, neurovascular, meniscal, ligamentous, and tendinous structures for injury or inflammation resulting from mass effect or impingement [20, 26, 30] (Figs. 4–6). By assisting planning for definitive resection while minimizing damage to more normal cartilage, MRI provides invaluable information regarding the presence and location of a cleft between the lesion and underlying normal epiphysis, which is generally seen as a linear margin of tissue with signal properties matching normal cartilage intervening between the lesion and underlying normal secondary ossification center or physis [31] (Figs. 1 and 4). Of note, this differs from osteochondromas, which show contiguity with their underlying cortex and follow a normal pattern of growth and endochondral ossification contiguous with the parent bone [4].

Whole-body MRI may have a role in screening for additional lesions without ionizing radiation exposure within the same setting as initial lesion characterization; however, the benefit of this approach should be weighed against its availability, cost, and whether sedation is required for a short-protocol whole-body MRI examination [32]. After resection,

MRI is the preferred means of assessing for any suspected residual or recurrent lesion [33].

Ultrasound

Ultrasound provides an incomplete evaluation of DEH lesions but can image cartilaginous overgrowth seen in the early stages of DEH before endochondral ossification (Fig. 2). It may be useful in suggesting DEH in the setting of a periarticular mass in a child before definitive evaluation with MRI.

Nuclear Medicine

Reflecting a primarily hypertrophic process, DEH lesions show increased uptake on bone scintigraphy [34]. However, the nonspecific nature of this radiotracer uptake precludes making definitive conclusions regarding DEH on bone scans. In addition, the availability of whole-body MRI to identify and characterize DEH lesions without radiation exposure generally obviates bone scintigraphy for children [20].

Natural History and Clinical Management

As the child with DEH grows, epiphyseal overgrowth advances with an increasing size discrepancy noted between the normal epiphysis side and the affected other side. The growth of these lesions is expected to continue until the child reaches skeletal maturity [5]. This size discrepancy frequently results in joint deformities, and DEH may result in limb length discrepancies (Fig. 2) and

gait disturbances, most often with hip or knee involvement [1, 35]. Enlarging osteocartilaginous lesions may cause progressive joint deformity, and DEH lesions may fragment as a result of mechanical stress with movement [3]. Functionally, large intraarticular lesions can result in early osteoarthritis in approximately 15% of cases [1]. Fortunately, no cases of malignant transformation of DEH lesions have been reported to date [1, 4].

Given the frequency of functional deformity, the risk of premature osteoarthritis, and the prevalence of pain, surgical resection of osteocartilaginous lesions is the preferred treatment method [1, 7]. Biopsy before resection is not generally indicated because of the highly characteristic nature of imaging features, but it may be helpful in unusual cases [1, 10, 36, 37]. Resection is generally definitive, resulting in good outcomes and very few reported instances of lesion recurrence [1, 26, 28]. Some have cautioned against resection of intraarticular lesions and others have endorsed nonoperative approaches for patients with asymptomatic DEH, but a review of published cases points out substantial clinical deterioration in a few conservatively managed cases, leading others to recommend intraarticular resection [1, 19, 30, 31]. Surgical approaches are determined on an individual basis and depend on the joint involved [7, 23, 35]. For example, acetabular involvement may particularly necessitate intraarticular lesion resection to preserve the hip joint, and DEH of the ankle may require arthrod-

esis [5, 23]. One study has reported cartilage surface-sparing wedge chondroplasty as an alternative [38]. Ultimately, no official consensus exists regarding the precise management of these lesions, although the prevailing practice is definitive resection combined with corrective osteotomies, when necessary, to avoid or correct growth discrepancies and deformities [1, 7, 23, 31, 35].

Related Conditions and Mimickers

Because of the overlap with several closely related disturbances of normal skeletal development, confusion regarding nomenclature, and the only more recent appreciation of differences between DEH lesions and osteochondromas, epiphyseal osteocartilaginous lesions in DEH may be mistaken for other conditions.

Osteochondromas

As previously mentioned, DEH lesions differ pathologically from osteochondromas, which are also much more frequent than DEH and affect older children and adults [15, 18]. Osteochondromas generally arise from the metaphyseal regions and grow away from the joint, differing significantly from DEH in anatomic location [4]. In addition, they exhibit continuity with the underlying nor-

mal-appearing medullary cavity, whereas osteocartilaginous lesions in DEH are generally separate from the underlying epiphyseal medullary cavity.

Hereditary osteochondromatoses, including multiple hereditary exostoses, typically present with bilateral involvement, which rarely occurs in DEH [4]. One particular subtype of osteochondromatosis, dominant carpotarsal osteochondromatosis, shares the presence of epiphyseal osteocartilaginous lesions in common with DEH and can be indistinguishable on imaging [39]. Dominant carpotarsal osteochondromatosis, however, occurs in a hereditary pattern with autosomal dominant inheritance and has bilateral involvement by definition [40, 41].

Metachondromatosis

Metachondromatosis is an infrequently encountered genetic disease of both osteochondroma and enchondroma formation attributed to the *PTPN11* gene [42]. Children with this condition may have periarticular calcified soft-tissue masses develop that may simulate the appearance of osteocartilaginous lesions seen in DEH. Of importance, these soft-tissue masses tend to occur in the hands near the phalanges, as in the example

shown in Figure 7. Metachondromatosis differs significantly in its anatomic distribution, favoring the hands and feet, with femoral, tibial, and pelvic lesions occurring less frequently. Moreover, the metaphyses are the primary site of involvement in metachondromatosis, often with exophytic osteochondroma or diffuse enchondroma lesions [42].

Multiple Epiphyseal Dysplasia

Multiple epiphyseal dysplasia (also called Fairbank disease) encompasses a collection of related hereditary disorders in which flattening and fragmentation of the epiphyses occur (Fig. 8). This skeletal dysplasia differs strikingly from DEH because of the smaller size of the affected epiphysis as opposed to the hypertrophy encountered in DEH [1]. Moreover, the anatomic distribution of multiple epiphyseal dysplasia is not expected to follow the patterns described in DEH. Of note, this condition also entails dwarfism that is not part of the clinical presentation of DEH.

Mimickers

A few entities can potentially mimic DEH, mostly on radiographic examination. Ossific densities seen in synovial chondromatosis (osteochondromatosis) may project in the
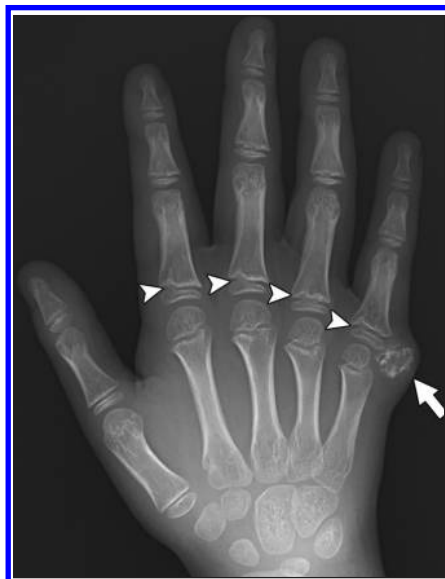

Fig. 7—9-year-old boy with palpable mass. Posteroanterior radiograph of right hand shows lobular calcified subcutaneous mass near right fifth metacarpophalangeal joint (*arrow*) as well as diffusely abnormal shape of metaphyses of proximal and middle phalanges and metacarpals of both hands, with large cartilage tongues extending into metaphyses of proximal phalanges (*arrowheads*). Taken together, these findings are most indicative of metachondromatosis, for which patient had genetic testing that confirmed diagnosis.
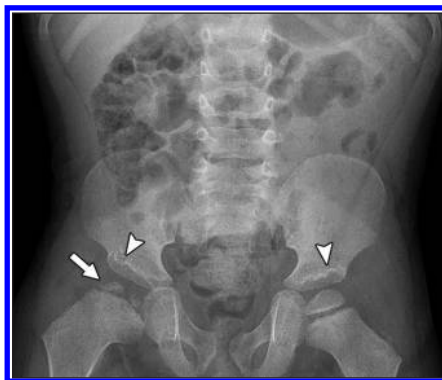
Fig. 8—3-year-old boy with suspected Legg-Calvés-Perthes disease.
A, Skeletal survey radiograph of pelvis shows multiple affected epiphyses with flattening of right proximal femoral epiphysis (*arrow*), irregularity of both acetabula (*arrowheads*), and flattening of multiple vertebral bodies. In addition, irregularity of metaphyses and epiphyses of multiple long bones bilaterally were also observed (not shown).
B, Follow-up radiograph of right ankle obtained when patient was 10 years old shows progressive flattening and irregularity around physes, particularly involving distal tibia (*arrow*). Diagnosis of multiple epiphyseal dysplasia was made on basis of imaging findings and was confirmed by genetic testing. These findings contrast with those of dysplasia epiphysealis hemimelica, in which epiphyseal overgrowth occurs rather than epiphyseal flattening, as was observed in this case.

Fig. 9—7-year-old boy with decreased range of motion in left elbow and history of prior elbow trauma. Frontal radiograph of left elbow shows deep concavity of trochlear groove (*arrowhead*) and disorganized ossific fragments in region of trochlear epiphysis (*arrow*). Premature partial closure of both trochlear and radial physeal regions is noted. Findings are consistent with fishtail posttraumatic skeletal growth abnormality resulting from development of osteonecrosis after injury.

Fig. 10—14-year-old girl who presented with chronic ankle pain.
A and **B**, Mortise view ankle radiograph (**A**) shows multiple discrete ossific densities around lateral aspect of tibiotalar joint and lateral gutter (*arrow*). Lateral ankle radiograph (**B**) shows additional ossific densities (*arrowheads*). These findings were confirmed to represent synovial chondromatosis on arthroscopic removal.

region of an epiphysis to simulate ossification centers (Fig. 9). However, synovial chondromatosis (osteochondromatosis) generally follows a broader distribution that is not confined to the epiphysis and occurs in an older population [3]. Similarly, posttraumatic arthrosis with multiple intraarticular bodies may be confused with DEH in the absence of a provided trauma history. Osteonecrosis of the distal humerus encountered in the so-called fishtail deformity (Fig. 10) of chronic posttraumatic skeletal growth abnormality can be particularly vexing because the clinical presentation of pain and decreased range of motion are often delayed, resulting in an unclear medical history at the time of imaging [43]. Nonetheless, careful inspection, especially on MRI, for the presence of epiphyseal overgrowth that would be expected in DEH only should exclude these mimickers from the differential diagnosis.

Conclusion

DEH (Trevor disease), although infrequent, has significant implications for the growing child. An understanding of the typical and unusual imaging manifestations of this condition is helpful for any radiologist evaluating children because it improves detection of epiphyseal growth abnormalities. The more recent pathologic literature sup-

ports this entity as a unique epiphyseal cartilage overgrowth distinct from osteochondromas. We propose a revised understanding of imaging findings in DEH as representing a range of abnormalities ranging from isolated epiphyseal hypertrophy to numerous ossification centers to metaphyseal involvement. Imaging is useful in guiding interventions to avert growth disturbance, pain, deformity, and early degenerative disease. Several imaging modalities provide complementary information regarding these lesions and clearly distinguish these osteocartilaginous lesions from other entities such as osteochondromas. MRI provides conclusive imaging of unossified lesions in DEH and allows detection of associated involvement of adjacent structures, including the epiphyseal and metaphyseal involvement that has been acknowledged more recently and was not included in original definitions based on radiographs.

Acknowledgment

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