

Pelvic radiograph in skeletal dysplasias: An approach

Manisha Jana, Nikhil Nair, Arun K Gupta, Madhulika Kabra¹, Neerja Gupta¹

Departments of Radiodiagnosis and ¹Pediatrics, All India Institute of Medical Sciences, New Delhi, India

Correspondence: Dr. Manisha Jana, Room No. 81C, Department of Radiodiagnosis, All India Institute of Medical Sciences, Ansari Nagar, New Delhi, India. E-mail: manishajana@gmail.com

Abstract

The bony pelvis is constituted by the ilium, ischium, pubis, and sacrum. The pelvic radiograph is an important component of the skeletal survey performed in suspected skeletal dysplasia. Most of the common skeletal dysplasias have either minor or major radiological abnormalities; hence, knowledge of the normal radiological appearance of bony pelvis is vital for recognizing the early signs of various skeletal dysplasias. This article discusses many common and some uncommon radiological findings on pelvic radiographs along with the specific dysplasia in which they are seen; common differential diagnostic considerations are also discussed.

Key words: Achondroplasia; pelvic radiograph; skeletal dysplasia

Introduction

The bony pelvis is constituted by ilium, ischium, pubis, and sacrum. The pelvic radiograph is commonly included in routine radiographic examinations, and consequently often serves as the first evidence of skeletal dysplasia. Knowledge of the normal pelvic embryological development is vital for recognizing the early signs of various skeletal dysplasias manifesting in pelvis. The pelvic radiograph often reveals characteristic abnormalities in pelvic shape, size, number of bones, density, timing of ossification, etc., which may provide vital clues in the recognition of various dysplasias. Caffey was the first to recognize the importance of the pelvis in the diagnosis of bone dysplasias when he conducted a detailed analysis of the skeletal anomalies encountered in achondroplasia.^[1] Further down the line, Kaufmann^[2] described many other patterns of malformation, which shed more light on the significance of pelvis in diagnosing

constitutional diseases of bone. This article discusses many common and some uncommon radiological findings on pelvic radiographs along with the specific dysplasia in which they are seen; common differential diagnostic considerations are also discussed.

Ossification

For interpretation of radiographic abnormalities in bony pelvis, thorough knowledge about the ossification centres in and around the pelvis is essential. The time of appearance of the ossification centres is listed in Table 1.

In the full-term neonatal pelvic radiograph, ilium, superior pubic, and ischial rami, 1st to 5th sacral vertebrae, 1st to 4th neural arches, 1st and 2nd lateral centres, and the first coccygeal vertebra are usually seen to be ossified [Figure 1].^[3]

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

Cite this article as: Jana M, Nair N, Gupta AK, Kabra M, Gupta N. Pelvic radiograph in skeletal dysplasias: An approach. Indian J Radiol Imaging 2017;27:187-99.

Access this article online

Quick Response Code:



Website:
www.ijri.org

DOI:
10.4103/ijri.IJRI_367_16

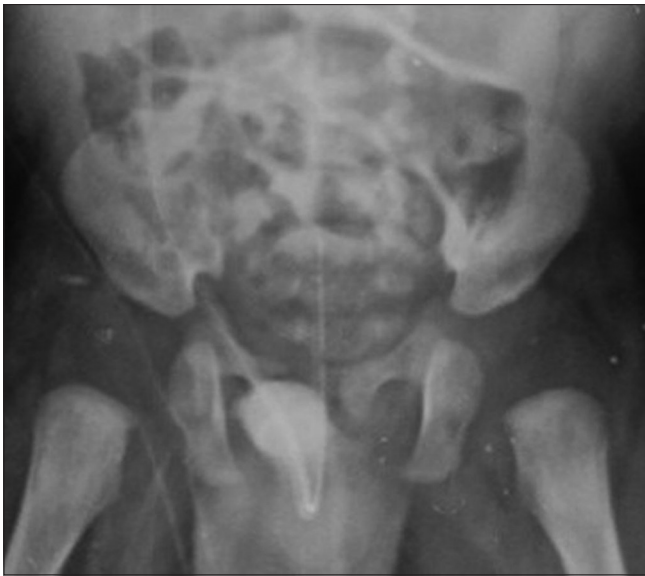


Figure 1: Normal pelvic radiograph in a full term newborn

Table 1: Appearance of ossification centres in bony pelvis

Bone	Appearance of primary ossification centre
Ilium	8 weeks in utero
Ischium	18 weeks in utero
Pubis	20 weeks in utero
Sacrum	14 weeks in utero
Acetabulum (two centres)	Puberty
Femoral head	4-6 months
Greater trochanter	2-4 years
Lesser trochanter	Puberty

Abnormalities in different skeletal dysplasias

Iliac bone and acetabulum

- **Flared iliac wing and flat acetabular roof** – Characteristic radiological signs seen in both achondroplasia and lethal dysplasias. The typical appearance in achondroplasia has been given numerous descriptions – “tombstone shape;” “mickey mouse ear pelvis;” “champagne glass pelvis.”
- **Square iliac wing** (vide supra) – Refers to small square-shaped iliac wings akin to a tombstone. This sign is seen in the most common skeletal dysplasia, i.e., achondroplasia. Incidentally, it is also seen in the second most common lethal skeletal dysplasia viz., thanatophoric dysplasia. Rhizomelic dwarfism is a common feature of both dysplasias; however, telephone handle shaped femur, short horizontal ribs, and cloverleaf skull are distinctive features of thanatophoric dysplasia.^[4] Similarly, champagne glass shaped pelvic inlet, trident hand and pelvis (described below), metaphyseal flaring and horizontal acetabular roof are some of the unique features of achondroplasia that enable easy differentiation between the two entities [Figure 2]
- **Hypoplastic iliac wing** – Seen in several spondyloepiphyseal dysplasias, namely

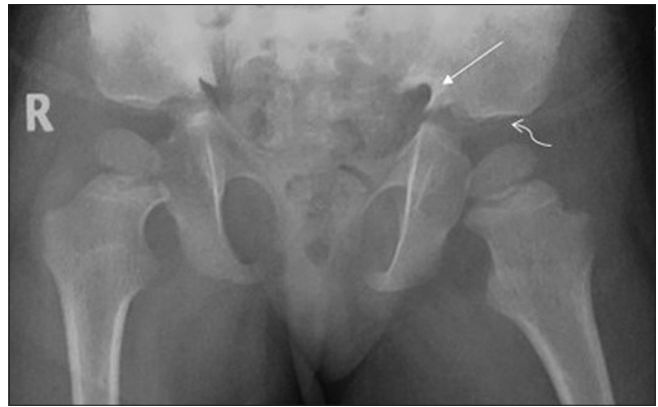


Figure 2: Pelvis radiograph in achondroplasia shows square shape of the iliac bone, horizontal acetabular roof (squiggly arrow), and rhizomelic shortening of the femur. Also note the trident sign in achondroplasia. The three-pronged pear shape of the sciatic notch is well visualized in this radiograph (straight arrow)

spondyloepiphyseal dysplasia tarda with mental retardation, spondylomegaepiphyseal dysplasia, and Dyggve–Melchior Claussen (DMC) syndrome. It should be differentiated from foreshortening of iliac bones in campomelic dysplasia (see below).

- **Champagne glass pelvis/mickey mouse ear pelvis** (also see above) – Refers to the flattening of iliac blades with increased acetabular angles and small sacrosiatic notch giving rise to a champagne glass shaped pelvic inlet. This sign is seen in achondroplasia [Figure 2].
- **Sloping acetabular roof** – Refers to shallow acetabular fossae and steep slope of acetabular roof, which is a common feature in mucopolysacchridosis. Similar finding may also be seen in acetabular dysplasia which may occur as a genetically determined condition or secondary to incomplete reduction of a congenital dislocation, damage to the lateral acetabular epiphysis, or femoral head maldevelopment [Figure 3].^[5]
- **“Trident sign”** – Spurs at the medial and lateral acetabular margin and in the center of the acetabulum gives rise to shape resembling a three-pronged spear known as trident [Figure 2]. This sign is seen in achondroplasia, Jeune’s asphyxiating thoracic dystrophy (JATD), Ellis-van Creveld syndrome, and thanatophoric dysplasia.^[6] Mesomelic pattern of dwarfism and bell shape of the chest are distinctive features of Ellis-van Creveld syndrome.

The imaging findings in achondroplasia include

- Trident pelvis/tombstone shape of the iliac bone
- Rhizomelic shortening of long bones with metaphyseal flaring
- Gradual narrowing of interpedicular distance in the lumbar spine
- Short pedicles of lumbar vertebrae
- Large calvarium and a relatively small base
- Trident hand (increased gap between third and fourth fingers)

JATD shows short ribs and a narrow thorax [Figure 4].

Thanatophoric dysplasia is a lethal dysplasia with *FGFR3* mutation (allelic but much severe than achondroplasia and hypochondroplasia) with characteristic “cloverleaf skull,” extreme long bone shortening, bowed appearance of long bones (telephone handle shape of femora), more severe rib shortening, and increased platyspondyly with characteristic U and H-shaped vertebrae.^[7]

Imaging clues towards Ellis-van Crevald syndrome (clinical setting of post-axial polydactyly, midline cleft lip and cardiac anomalies) include

- Trident pelvis
- Acro-mesomelic limb shortening
- Outward bowing of humeri
- Bell-shaped thorax improving with age
- Laterally deficient upper tibial epiphyses
- Cone shaped epiphyses in hands, capito-hamate coalition
- **Foreshortened iliac bone** – In campomelic dysplasia, the iliac bone is small, oriented in the anterior-posterior direction, and hence appears foreshortened [Figure 5]. Campomelic dysplasia is a lethal dysplasia associated with XY sex reversal.

This appearance is also seen in chondroectodermal dysplasia and cleidocranial dysostosis. Bowed long bones, hypoplastic scapulae, and narrow chest seen in campomelic dysplasia allow differentiation from these entities.

The imaging pointers towards campomelic dysplasia include

- Bowing of long bones of extremities
- Angulated bowing of femur at the junction of upper third and middle third
- Angulated bowing of tibia at the middle and lower third junction
- Small/foreshortened iliac wing
- Hypoplastic angle of scapula
- **Lateral acetabular notch** – Refers to a notch at the superolateral margin of the acetabulum seen in acromesomelic dysplasia and in some cases of neonatal hip dysplasia
- **Lacy iliac wing** – This sign is ascribed to the iliac crest irregularity due to dystrophic ossification of the osteochondral junction giving an appearance of a lace border around the iliac crest. This radiological sign is a pathognomonic feature of Dyggve- Melchior Claussen (DMC) syndrome [Figure 6].^[8] Other pelvic findings in this syndrome include short iliac wings, wide sacroiliac joints, narrow sciatic notches, wide pubic and ischial rami, dysplastic acetabular fossae, and laterally displaced femoral heads [Figure 6]. Similar findings are seen in enchondromatosis (Ollier’s disease) [Figure 7] as

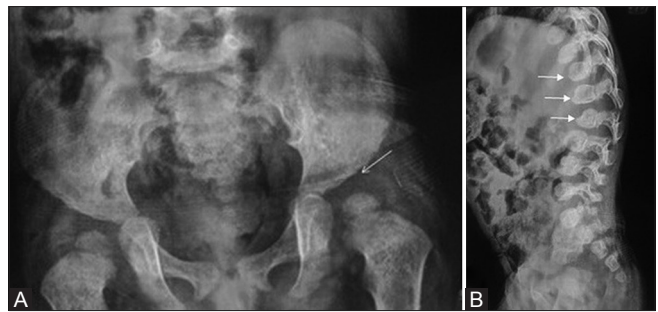


Figure 3 (A and B): (A and B) Pelvis radiograph in mucopolysaccharidosis type I (A) reveals steep sloping of bilateral acetabular roofs (marked), which are irregular. Lumbosacral spine radiograph lateral view (B) of the same patient reveals localized angular kyphosis at the dorsolumbar junction and beak-like projection (arrows) from the inferior aspect of visualized thoracolumbar vertebral bodies (inferior beaking)

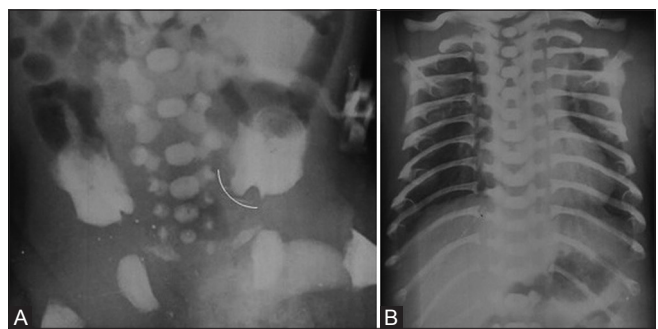


Figure 4 (A and B): Trident pelvis in Jeune’s asphyxiating thoracic dystrophy. Pelvis radiograph (A) shows a flared iliac bone having a trident shape (marked) at sciatic notch. The chest radiograph (B) reveals that the ribs are small and thoracic cavity narrow

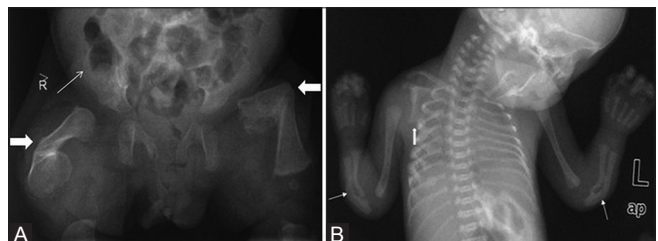


Figure 5 (A and B): (A, B) Campomelic dysplasia. Radiograph of pelvis (A) of the stillborn fetus shows apparent shortening of the iliac bone (thin arrow); and curved long bone of the lower limb (block arrow). Chest radiograph (B) revealed hypoplastic scapula (thick arrow) and bent radii and ulnae (thin arrows)

well as in Smith-McCort syndrome, a condition clinically and radiologically identical to DMC syndrome except for absence of mental retardation.^[9]

The imaging pointers towards DMC and Smith-McCort syndrome include

- Lacy iliac crest
- Dysplastic acetabulum with “laterally placed” femoral heads
- Wide sacroiliac joints
- Wide pubic and ischial rami

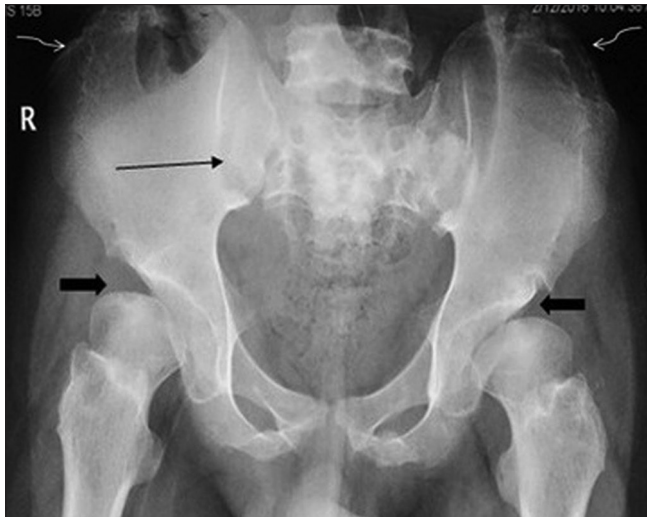


Figure 6: Dyggve-Melchior Claussen syndrome: Pelvic X-ray demonstrates bilateral iliac crest irregularity (squiggly arrows), wide sacroiliac joints (straight arrow), wide pubic, and ischial rami. Note dysplastic acetabular fossae with abnormally shaped and laterally displaced femoral heads (block arrows)

- Central notching in the vertebrae
- Lace-like appearance of scapular angle

Lacy appearance of iliac wings may also be seen in parastremmatic dwarfism (Greek word parastremmatic = twisted). Symmetrical bowing of the long bones, severe genu valgum, short neck, kyphoscoliosis, etc., permit easy differentiation of this entity from others.

- **“Paraglider” or “crescent” shape of iliac wing** – Deficient mineralization of the iliac wings in achondrogenesis gives rise to the characteristic “paraglider” shape of the iliac wings [Figure 8]. Achondrogenesis is a lethal skeletal dysplasia, with poor mineralization of the skeleton. Three types have been described, type IA, IB, and II. The imaging findings and the disease severity vary in these types. All the three types show a crescent/paraglider shape iliac bone. Type IA is a severe form of the disease, where the skull vault appears completely unmineralized. There is severe shortening of the long bones and the ribs are short and beaded. The vertebral bodies show poor mineralization (c.f: pedicles of thoracic vertebrae unmineralized in Campomelic dysplasia). In type IB, there is a typical expansion of the bony vertebral column at the lumbar level, giving rise to the “cobra head” appearance.
- **Halberd pelvis** – Metatropic dysplasia is a spondyloepimetaphyseal dysplasia with diagnostic radiographic findings comprising of marked platyspondyly, short tubular bones with broad metaphyses, and small epiphyses.^[10] The unique shape of the iliac wing – a notch between the shortened iliac body and prominent crescent-shaped iliac



Figure 7: Ollier's disease. Pelvic radiograph shows the characteristic lace-like irregular configuration of the iliac crest margins on both sides (marked). Also of note are the multiple enchondromas in bilateral proximal femoral shafts (marked on left side) and right iliac wing, which point towards the right diagnosis

wing resembles a two-handed pole weapon known as halberd. This pathognomonic radiological sign distinguishes this entity from other common forms of spondylometaphyseal and spondyloepimetaphyseal dysplasias.

- **Snail-like appearance of iliac wing** – Refers to hypoplastic iliac bones with a snail like configuration [Figure 9]. Seen in Schneckenbecken dysplasia (German for snail pelvis), a rare lethal neonatal chondrodysplasia with autosomal recessive inheritance.^[6] Other distinctive features seen in this disorder include flattened and hypoplastic vertebral bodies, short ribs, dumb bell-like appearance of long bones, and precocious ossification of the tarsal bones.^[6]
- **Sunburst appearance of iliac wings** – Dense bands of bone radiating from the acetabulum to iliac crest [Figure 10] in a fan-like pattern give rise to “sunburst” appearance which is a characteristic feature of osteopathia striata.^[11] Similar bands extending from metaphysis to diaphysis of long bones are the other important radiological feature of this dysplasia.
- **Posterior iliac horn** – Bilateral symmetrical posterior iliac horns [Figure 11] are a unique feature of Nail Patella syndrome (also known as Fong's disease). The horns are formed as a result of exostoses from posterior aspect of iliac bones and are considered as a pathognomonic feature seen in as many as 80% of the patients with this syndrome.^[12]
- **Bone within bone appearance** – Increased density of pelvic bones with pelvic endobones (bone within bone)



Figure 8: Representative illustration depicting relatively well ossified iliac bone with long crescent-shaped medial and inferior margins (arrows) typical of Paraglider shaped pelvis – classical finding seen in achondrogenesis



Figure 9: Representative illustration depicting extremely small, snail-shaped ilium (left) with a superomedial extension (arrow) resembling a snail's head – seen in Schneckbecken dysplasia



Figure 10: Osteopathia striata. Radiograph of the pelvis showing alternating sclerotic bands in the iliac wings (marked)

appearance is a classical feature of type II autosomal dominant osteopetrosis [Figure 12].^[13]

- **Multiple scattered sclerotic foci in pelvic bones:** Seen in osteopoikilosis, a sclerosing bony dysplasia characterised by multiple bone islands. The pelvis is a common site of involvement with clustering of sclerotic foci around hip and sacroiliac joints [Figure 13].

Sacrum and lower lumbar vertebrae

- **Calcific Stippling of sacrum and lumbar vertebrae** – Seen in chondrodysplasia punctata, especially Sheffield type. This disorder is characterized by calcific stippling of cartilage, particularly epiphyses of long bones.^[14] Stippling with rhizomelic shortening can suggest rhizomelic chondrodysplasia punctata [Figure 14].



Figure 11: Representative illustration depicting typical triangle-shaped excrescence arising from the posterior aspect of the ilium (posterior iliac horn)– classical finding seen in Nail–patella syndrome

The imaging findings include

- Stippling of sacral and lumbar vertebrae
- Rhizomelic shortening of long bones
- Tarsal bone stippling
- Coronal cleft vertebrae
- Scoliosis
- **Decreasing interpedicular distance in lumbar vertebrae** – seen in achondroplasia (see above)



Figure 12: Osteopetrosis. Radiograph of the pelvis reveals generalized sclerosis of the pelvic bones with loss of corticomedullary differentiation



Figure 13: Osteopoikilosis. Radiograph of the pelvis reveals multiple well defined sclerotic lesions around the pelvis arrows, especially centred around the joints

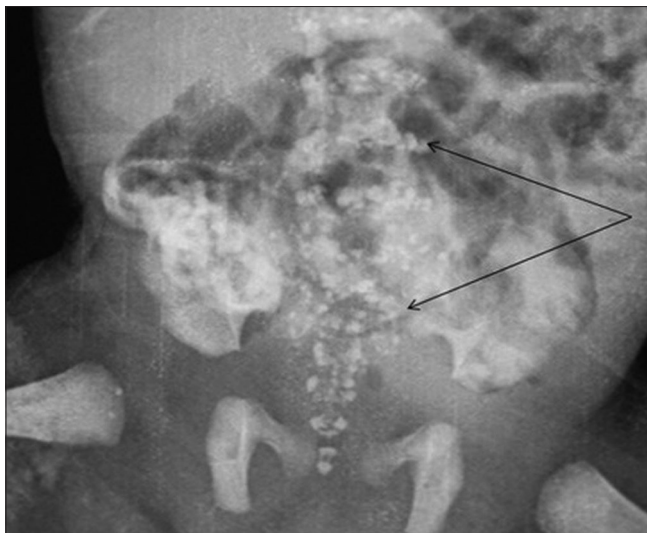


Figure 14: Chondrodysplasia punctata in another newborn. Pelvic radiograph shows extensive calcific stippling of sacrum and lower lumbar vertebrae (marked)

- **Medially placed, more rounded pedicles of lumbar vertebrae** – Seen in spondylometaphyseal dysplasia Kozlowski type.
- **Absent sacral vertebrae** – Aplasia/dysplasia of the lumbosacral vertebral bodies is a common feature of caudal regression syndrome, sometimes associated with complete sacral agenesis [Figure 15].

Pubic bone and triradiate cartilage

- **Absent ossification of the pubic bone** – Among the pathognomonic features of cleidocranial dysostosis is retarded/absent ossification of the pubic bones which gives rise to an apparent increase in interpubic distance (pseudo diastasis) [Figure 16]. This feature is also seen in achondrogenesis, hypochondrogenesis (short long bones), Sjogren Larssen syndrome, Wolf syndrome, and spondyloepiphyseal dysplasia congenital.^[15]



Figure 15 (A and B): Caudal regression syndrome. Lumbosacral spine AP radiograph (A) in a neonate shows absence of lower sacral vertebrae and coccyx. Lateral projection of lumbosacral spine in same patient shows presence of L1 vertebra (marked) with absence of remaining lower vertebrae

Imaging features of cleidocranial dysostosis include

- Widely spaced pubic bones, pubic diastasis like appearance
- Elongated femoral head (chef's hat sign)
- Accessory epiphyses at the second metacarpal base

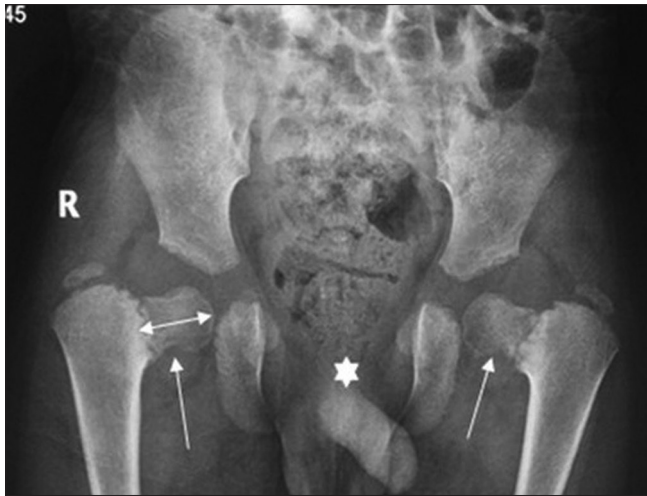


Figure 16: Cleidocranial dysostosis. Pelvic radiograph revealing retarded ossification of pubic bones resulting in widening of symphysis pubis (pseudo diastasis, asterisk). Also note the elongated shape of capital femoral epiphyses giving a “chef’s hat” sign, along with coxa vara (arrows)

- Wide open cranial sutures
 - Multiple wormian bones
 - Multiple unerupted teeth
 - Distal phalangeal tapering
 - Hypoplastic/absent clavicles
- **True dislocation of hip** – Superolateral displacement of femoral head and shallow acetabulum is seen in the developmental dysplasia of hip. Other important pelvic radiographic findings include small capital femoral epiphysis, delayed femoral head ossification, and acetabular sclerosis. Delayed femoral head ossification renders plain film radiography unreliable in children less than 12 months of age with ultrasonography being the optimum imaging modality.
 - **Apparent dislocation of hip** – Delayed ossification of capital femoral epiphyses and horizontal acetabular roofs result in this radiographic appearance seen in spondyloepiphyseal dysplasia congenita (SEDC). This disease is a type II collagenopathy, having autosomal dominant transmission, and clinical presentations include hearing loss and retinal detachment. Other important pelvic findings in this disorder include metaphyseal flaring, triangular fragment on the inferior aspect of the broad femoral neck, and coxa vara [Figure 17]. Similar findings can also be seen in spondyloepimetaphyseal dysplasia (Strudwick type) [Figure 18], but with more prominent metaphyseal irregularity.

SEDC has the imaging features as follows

- Absent ossification of pubic bones
- Apparent dislocation of hip joints
- Coxa vara

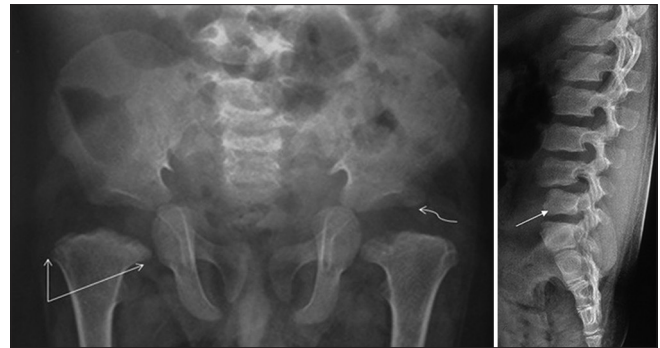


Figure 17 (A and B): SEDC with pelvic radiograph (A) demonstrating delayed ossification of bilateral femoral heads and horizontal configuration of acetabular roofs (squiggly arrow). Also note bilateral femoral metaphyseal flaring (straight arrows) and coxa vara. Radiograph of lumbosacral spine (B) in the same patient depicts anisospondyly (L4 vertebral body larger than L5 vertebral body) (arrow)

- Poorly developed femoral head epiphyses
- Platypondyly
- Anisospondyly (variable size of lumbar vertebrae; e.g., L1 bigger than L5)

Spondyloepiphyseal dysplasia tarda (SEDT), in comparison, has a typical morphology of the vertebral bodies; a “hump” at the posterior two-thirds of the vertebral bodies.

- **Pubic diastasis** – Refers to abnormally wide gap between the pubic bones of >10 mm. This radiological sign is seen in Bladder exstrophy, Prune Belly syndrome, and osteogenesis imperfecta.
- **Wide triradiate cartilage** – Widening and delayed fusion of triradiate cartilage is seen in pseudoachondroplasia [Figure 19].

The imaging findings in pseudoachondroplasia are

- Square iliac bones (similar to achondroplasia)
- Wide triradiate cartilage
- Delayed ossification of femoral head
- Metaphyseal flaring
- Irregular epiphyses
- Tongue like projection from anterior aspect of vertebrae
- Widened costo-chondral and costo-transverse junction.

Proximal femur and hip joints

- **Coxa vara** – Refers to characteristic hip deformity due to angle between femoral neck and shaft being <120 degrees. This sign is nonspecific and can be seen in many skeletal dysplasias such as spondyloepiphyseal dysplasia congenita, spondyloepimetaphyseal dysplasia Strudwick type, congenital coxa vara with Fairbank’s triangle, and pseudoachondroplasia.
- **Coxa valga** – Refers to abnormally increased femoral neck shaft angle which may range normally up to 160 degrees in newborns and 145 degrees in children. This



Figure 18: SEMD: Pelvic radiograph reveals imaging findings similar to SEDC, with metaphyseal flaring and coxa vara – note the prominent metaphyseal irregularity (arrow)



Figure 19: Pelvic radiograph in pseudoachondroplasia in a 10-year-old reveals wide triradiate cartilage (arrows) and small femoral head for age (block arrow)

sign is seen in mucopolysaccharidosis [Figure 3], Turner syndrome, and developmental dysplasia of hip.

- **Small capital femoral epiphyses/irregularity and fragmentation/snow cap epiphysis** – Seen in Perthes' disease/avascular necrosis of head of femur. If bilateral, search should be made for any history of steroid intake, hypothyroidism, sickle cell disease, and other hemoglobinopathy. ARMED [Figure 20] is an autosomal recessive form of multiple epiphyseal dysplasia which can mimic the appearance of bilateral Perthes' disease.

The imaging findings in multiple epiphyseal dysplasia include

- Flattened irregular capital femoral epiphyses
- Subluxation of femoral head/protrusio acetabulae
- Cone-shaped epiphyses
- Irregular carpal bones
- Delayed bone age

Double-layered patella, a characteristic finding seen in the lateral knee radiograph, is found in ARMED.

- **Chef's hat sign** – Attributed to deformed capital femoral ossification centre with a rounded appearance and tapered edge directed towards femoral neck which gives the overall impression of a "Chef's Hat."^[16] This sign is a characteristic feature of cleidocranial dysplasia with absent ossification of the pubic symphysis being the other important pelvic sign of this dysplasia [Figure 16].
- **Prominent medial beak at medial femoral neck** – Characteristic feature of pseudoachondroplasia, a rare rhizomelic short limbed skeletal dysplasia which shows radiographic similarity with achondroplasia in many aspects.^[17] However, unlike achondroplasia, the skull is normal with normal interpedicular distance and marked platyspondyly [Figure 21].^[17]

- **"Monkey-wrench femora"** – Seen in two rare osteochondrodysplasias, i.e., diastrophic dysplasia/desboquis dysplasia and Larsen's syndrome of Reunion Island. In diastrophic dysplasia, the typical appearance of the femoral heads resembling a Swedish Key or monkey wrench [Figure 22], is a pathognomonic radiological sign with other important radiological features including advanced carpal ossification and radial deviation of the second phalanx with a supernumerary ossicle at base of second phalanx.^[18] Larsen's syndrome of Reunion Island shows similar clinical and radiological appearances including monkey wrench femora (inconstant feature), however, extra ossification centre is not seen in these patients enabling differentiation between the two.^[18]
- **Irregular metaphyses of femur** – Seen in nutritional rickets, vitamin D dependent rickets, metaphyseal dysplasias, and spondylometaphyseal dysplasias. The different types of metaphyseal dysplasia are Schmidt type, McKusick type [Figure 23], and Jansen type (although several other rarer types exist). Spondylometaphyseal dysplasia group of diseases have additional changes in the spine.
- **"Corner fracture" at upper femoral metaphysis** – Unique radiological feature seen in spondylometaphyseal dysplasia Sutcliffe type.^[19] Spondylometaphyseal dysplasia Sutcliffe type (corner fracture type) is a rare disorder involving the spine and the long bone metaphyses. The clinical presentation is with waddling gait and coxa vara with imaging revealing distinctive "corner fractures" at the ends of long bone metaphyses [Figure 24]. Bilateral coxa vara is another important pelvic finding in this disorder. Additional imaging features include irregularity at vertebral endplates, odontoid hypoplasia, and ovoid shape of vertebral bodies. The differential diagnosis of "corner fractures" includes nonaccidental injury of childhood,

which needs to be carefully ruled out. Points favoring spondylometaphyseal dysplasia will include the presence of vertebral changes, generalized metaphyseal irregularity, and lack of any other fractures in the body.^[19]

- **Delayed ossification and fusion of femoral head** – Feature of Meyer’s dysplasia and Perthes’ disease. Other differential diagnoses include SEDC, pseudoachondroplasia, multiple epiphyseal dysplasia, hypothyroidism [Figure 25], dyschondroplasia, arthritis, and infection.



Figure 20: Small capital femoral epiphyses in multiple epiphyseal dysplasia. Radiograph of the pelvis shows small and irregular bilateral femoral head (marked); akin to bilateral Perthe’s disease. Radiograph of the knee (not shown here) showed irregular epiphyses around the knee joint, and a tibiotalar slant was apparent in ankle radiograph

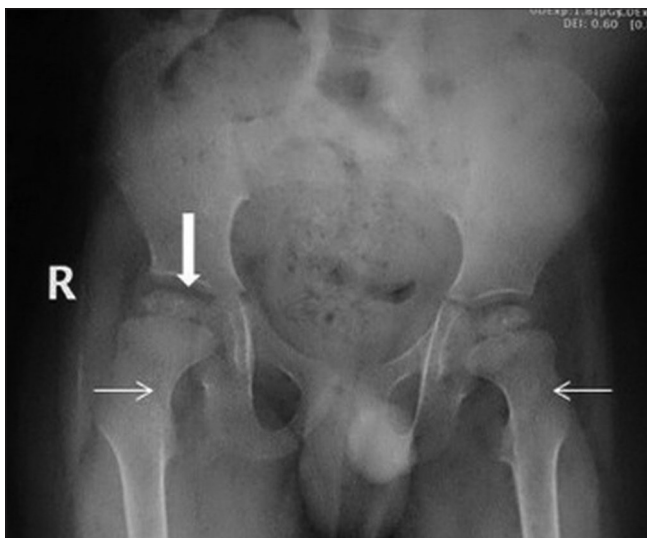


Figure 22: Case of Desboquis dysplasia – Anteroposterior radiograph of the pelvis showing “monkey wrench” appearance of both femora, elevated greater trochanters (thin arrows), coxa vara and dysplastic femoral epiphyses (thick arrow)

The imaging clues which can support a diagnosis of juvenile or congenital hypothyroidism are

- Delayed bone age
- Small irregular epiphyses
- Irregular sclerosis of metaphyses
- Wide triradiate cartilage
- Dorsolumbar kyphosis, bullet-shaped L1 vertebra
- Enlarged sella
- **Delayed ossification of femoral head** – Seen in Mucopolysacchridosis, pseudoachondroplasia, spondyloepiphyseal dysplasia, and developmental dysplasia of the hip. Irregular ossification of the femoral head in mucopolysacchridosis along with presence of bilateral coxa vara and bulbous femoral metaphyses in pseudoachondroplasia^[20] helps in differentiating between these entities based on pelvic radiographic findings.

Abnormal soft tissue ossification

Seen in fibrodysplasia ossificans progressiva [Figure 26], a rare hereditary mesodermal disorder with pelvic radiographs



Figure 21 (A and B): Pseudoachondroplasia – Pelvic radiograph (A) demonstrating metaphyseal flaring (block arrows), squared iliac wings, and horizontal acetabular roofs (squiggly arrow) akin to achondroplasia. Note that the femoral head are small (straight thin arrow). Also, in lateral spine radiograph (B), the tongue-shaped projection from the lumbar vertebrae (arrows) are a specific imaging finding in pseudoachondroplasia



Figure 23: Irregular upper femoral metaphyses in McKusick type metaphyseal chondrodysplasia

revealing ectopic soft tissue ossification which may ultimately result in bony bridging between thorax and pelvis.^[21]

The imaging findings in this rare condition include

- Multiple sheet-like ossification in soft tissues
- Bridging ossifications in soft tissues of neck and back
- Hallux valgus
- Microdactyly of the great toe

Conclusion

Radiograph of the pelvis is an important component of the skeletal survey in suspected skeletal dysplasia. Even

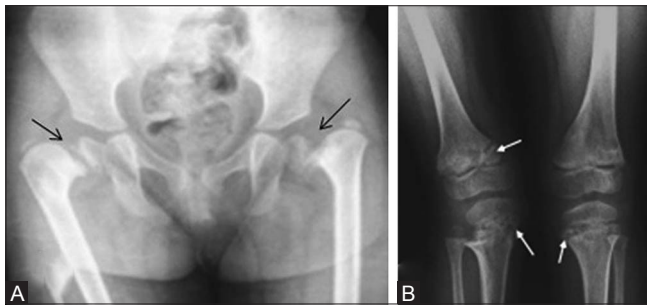


Figure 24 (A and B): Spondylometaphyseal dysplasia Sutcliffe type – (A) Pelvis with hip joint anteroposterior radiograph reveals bilateral coxa vara and slipped capital femoral epiphyses (B) bilateral knee anteroposterior radiograph reveals metaphyseal fraying with corner fractures (arrows)

in isolation, it can provide a host of information to guide the radiologist towards a possible group of disorders; thus, helping to perform a more specific/targeted search for additional findings to finally reach a diagnosis.

Essentials

- Knowledge of the ossification centres in the bony pelvis is very important [Table 1]



Figure 25: Juvenile hypothyroidism – Pelvic radiograph revealing unfused femoral head epiphysis and apophysis (small arrow) with persistent triradiate cartilage (arrows) and irregular epiphyses (block arrows)

Table 2: Differential diagnoses of various imaging findings in pelvis radiograph

Imaging findings	Differential diagnoses
Square iliac wing	Achondroplasia, Thanatophoric dysplasia
Trident pelvis	Achondroplasia
	Jeune Asphyxiating Thoracic Dysplasia
	Ellis van Creveld syndrome
	Thanatophoric dysplasia
Lacy iliac crest	DyggveMelchior-Clausen syndrome
	Smith-McCort syndrome
	Enchondromatosis, Parastremmatic dwarfism
Small/foreshortened iliac wing	Campomelic dysplasia
	Cleidocranial dysostosis
	Chondroectodermal dysplasia
Crescent shaped iliac wing	Achondrogenesis
Lateral acetabular notch	Acromesomelic dysplasia
Pubic diastasis/small pubic bones	Cleidocranial dysostosis
	Spondyloepiphyseal dysplasia congenita
	Achondrogenesis
Apparent dislocation of hip	Spondyloepiphyseal dysplasia congenita
Small femoral head	Spondyloepiphyseal dysplasia congenita
	Pseudoachondroplasia
Irregular flattened femoral head	Perthes disease, Multiple Epiphyseal Dysplasia, Spondyloepiphyseal dysplasia tarda
Delayed appearance and fusion of femoral head epiphyses	Juvenile hypothyroidism
	Pseudoachondroplasia
	Spondyloepiphyseal dysplasia congenita, Multiple Epiphyseal Dysplasia
Corner fracture at upper femora	Spondylometaphyseal dysplasia (Sutcliffe type)
Calcific stippling of the sacrum and epiphyses around hip joint	Chondrodysplasia punctata
	Rhizomelic Chondrodysplasia punctata

Table 3: Concise approach of combining findings on pelvic radiograph; along with other pointers towards a definitive diagnoses

Imaging findings in pelvis	Spine	Chest	Long bones	Skull	Hands	Other findings	Diagnoses
<i>Square shaped iliac bone</i> <i>Trident shape of iliac bone</i> <i>Trident pelvis</i>	Decreasing interpedicular distance in the lumbar spine	Normal	Rhizomelic shortening, Metaphyseal flaring	Large calvarium, narrow skull base	Trident hand		Achondroplasia
<i>Trident pelvis</i>	Normal	Long narrow thorax, short ribs					Jeune Apshyxiating Thoracic Dysplasia
<i>Trident pelvis</i>	Normal	Bell shaped thorax	Acromesomelic shortening Outward bowing of humeri		Polydactyly Carpal coalition Cone shaped epiphyses	Cleft lip, cardiac defects	Ellis van Creveld Syndrome
<i>Trident pelvis</i>	Severe platyspondyly	Bell shaped thorax	Bent femora (telephone receiver)	Clover leaf skull		Lethal dysplasia	Thanatophoric dysplasia
<i>Sloping acetabular roof</i>	Antero-inferior beaking of vertebral body	Paddle shaped ribs	Metaphyseal irregularity	J shaped sella	Proximal pointing of metacarpals	Coxa valga Kyphosis (esp cervical); odontoid hypoplasia	Mucopolysacchridosis (type I)
<i>Sloping acetabular roof</i>	Central beaking of vertebral body	Paddle shaped ribs	Metaphyseal irregularity	J shaped sella	Proximal pointing of metacarpals	Coxa valga Kyphosis (esp cervical); odontoid hypoplasia	Mucopolysacchridosis (type IV)
<i>Small iliac wing</i>	Thoracic vertebral pedicles appear unmineralized	Hypoplastic scapulae Hook shaped clavicle	Bent bones	Normal		Lethal, XY sex reversal	Campomelic dysplasia
<i>Small iliac wing (crescent shaped)</i>	Unmineralized vertebral bodies	Beaded ribs	Poor mineralization	Poor mineralization of skull vault		Lethal dysplasia	Achondrogenesis
<i>Lacy iliac wing</i>	Irregular notching of endplates	Lacy appearance of scapular angle	Dysplastic acetabulum with "laterally placed" femoral heads	Normal	Normal		Dyggve-Melchior-Clausen syndrome (with intellectual disability) Smith McCort syndrome (without intellectual disability)
<i>Non visualization of pubic bone</i>	Unmineralized vertebral bodies	Beaded ribs	Poor mineralization	Poor mineralization of skull vault		Lethal dysplasia	Achondrogenesis
<i>Non visualization of pubic bone (pseudo-diastasis)</i> <i>"Chef's hat" appearance of femoral head</i>	Normal	Hypoplastic/absent clavicles	Normal	Wormian bones, open fontanelles, Multiple unerupted teeth	Accessory epiphyses at metacarpals; Acro-osteolysis		Cleido-cranial dysostosis
<i>Non visualization of pubic bone</i> <i>Apparent dislocation of hip joints</i>	Anisodondyly	Normal	Irregular/poor ossification of epiphyses	Normal	Abnormal epiphyses at wrist	Coxa vara	Spondyloepiphyseal dysplasia congenita
<i>Small and irregular femoral capital epiphyses</i>	Mild endplate irregularity; platyspondyly	Normal	Epiphyseal irregularity	Normal	Irregular epiphyses at wrist Cone shaped epiphyses	Tibiotalar slant in ankle Double layered patella in ARMED	Multiple epiphyseal dysplasia
<i>Delayed bone age at upper femoral epiphyses</i>	Anisodondyly	Normal	Irregular/poor ossification of epiphyses	Normal	Irregular epiphyses at wrist	Coxa vara	Spondyloepiphyseal dysplasia congenita
<i>Delayed bone age at upper femoral epiphyses</i>	Anisodondyly	Normal	Irregular/poor ossification of epiphyses Metaphyseal irregularity	Normal	Irregular epiphyses at wrist		Spondyloepimetaphyseal dysplasia (Strudwick type)

Contd...

Table 3: Contd...

Imaging findings in pelvis	Spine	Chest	Long bones	Skull	Hands	Other findings	Diagnoses
Delayed bone age at upper femoral epiphyses	Central "tongue shaped" projection at dorso-lumbar vertebrae	Prominent costo-vertebral junctions	Metaphyseal and epiphyseal irregularity	Normal	Metaphyseal and epiphyseal irregularity		Pseudoachondroplasia
Wide triradiate cartilage	Central "tongue shaped" projection at dorso-lumbar vertebrae	Prominent costo-vertebral junctions	Metaphyseal and epiphyseal irregularity	Normal	Metaphyseal and epiphyseal irregularity		Pseudoachondroplasia
Wide triradiate cartilage	Bullet shaped vertebra at dorsolumbar junction	Normal	Metaphyseal irregularity and sclerosis, Epiphyseal irregularity	Widened sella	Metaphyseal irregularity		Hypothyroidism



Figure 26: Pelvic radiograph in a case of fibrodysplasia ossificans progresiva reveals soft tissue ossification in bilateral thighs adjacent to femur (straight arrows) as well as in right hemipelvis adjacent to right iliac wing (squiggly arrow)

- Few named signs and specific morphology can help make a definitive diagnosis in some dysplasias [Table 2]
- A combined interpretation of the entire skeletal survey is essential to reach a diagnosis [Table 3].

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

- Caffey J. Achondroplasia of pelvis and lumbo-sacral spine: Some roentgenographic features. *AJR* 1958;80:449-57.
- Kaufmann HJ. Röntgenbefunde am kindlichen becken bei angeborenen skelettaffektionen und chromosomalen aberrationen. Stuttgart, Germany: Thieme; 1964.
- Kohler A, Zimmer EA. Borderlands of the normal and early pathologic in skeletal roentgenology. Case JT. 10th ed. New York: Grune and Stratton; 1956.
- Sharma M, Jyoti, Jain R, Devendra. Thanatophoric Dysplasia: A Case Report. *J Clin Diagn Res* 2015;9:QD01-3.
- Solomon L, Warwick DJ, Nayagam S. Apley's Concise System of Orthopaedics and Fractures, Third Edition. Boca Raton: CRC Press, 2005. 209. ePDF
- Eich GF, Babyn P, Giedion A. Pediatric Pelvis: Radiographic Appearance in Various Congenital Disorders. *Radiographics* 1992;12:467-84.
- Reid J, Davros W, Paladin A, Lee E, Carrico C. Pediatric Radiology. Oxford: Oxford University Press; 2013. pp. 305.
- Aglan MS, Temtamy SA, Fateen E, Ashour AM, Eldeeb K, Hosny GA. Dyggve–Melchior–Clausen syndrome: Clinical, genetic, and radiological study of 15 Egyptian patients from nine unrelated families. *J Child Orthop* 2009;3:451-8.
- Gupta V, Kohli A and Dewan V. Dyggve–Melchior–Clausen syndrome. *Indian Pediatr* 2010;47:973-5.
- Sulko J, Marik I, Wisniewska M, Zemkova D, Kozlowski K. Metatropic dysplasia – case reports. *Pol J Radiol* 2008;73:81-4.
- Roche CJ, O'Keefe DP, Lee WK, Duddalwar VA, Torreggiani WC, Curtis JM. Selections from the Buffet of Food Signs in Radiology. *Radiographics* 2002;22:1369-84.
- West JA, Louis TH. Radiographic findings in the nail-patella syndrome. *Proceedings (Baylor University Medical Center)* 2015;28:334-6.
- Sit C, Agrawal K, Fogelman I, Gnanasegaran G. Osteopetrosis: Radiological & Radionuclide Imaging. *Indian J Nucl Med* 2015;30:55-8.
- Sanfilippo A, Bartoletti S. Brachytelephalangic chondrodysplasia punctata: A difficult diagnosis. *Radiol Case Rep* 2010;51:Article 308.
- Cortina H, Vallcanera A, Andres V, Gracia A, Aparici R, Mari A. The non-ossified pubis. *Paediatr Radiol* 1979;8:87-92.
- Aktas S, Wheeler D, Sussman MD. The 'chef's hat' appearance of the femoral head in cleidocranial dysplasia. *J Bone Joint Surg Br* 2000;82:404-8.
- Tandon A, Bhargava SK, Goel S, Bhatt S. Pseudoachondroplasia: A rare cause of rhizomelic dwarfism. *Indian J Orthop* 2008;42:477-9.

18. Al Kaissi A, Radler C, Klaushofer K, Grill F. Advanced ossification of the carpal bones, and monkey wrench appearance of the femora, features suggestive of a probable mild form of desbequious dysplasia: A case report and review of the literature. *Cases J* 2009;2:45.
19. Nair N, Satapathy, Gupta N, Kabra M, AK Gupta, Jana M. Spondylometaphyseal Dysplasia Corner Fracture (Sutcliffe) Type. *Indian J Pediatr* 2016;83:1191-4.
20. Hefti F. *Pediatric Orthopedics in Practice* 2nd edition. Hiedelberg: Springer-Verlag; 2015. pp. 767.ebook
21. Al-Salmi I, Raniga S, Hadidi AA. Fibrodysplasia Ossificans Progressiva-Radiological Findings: A Case Report. *Oman Med J* 2014;29:368-70.