

Role of Computed Tomography Imaging in evaluation of pediatric congenital and developmental skeleton disorder

ABSTRACT

Introduction: Pediatric congenital and developmental disorder is common nowadays. That particularly occurs due to error in normal development of organ and tissue development. Genetic disorders affecting skeleton comprise a large group of clinically distinct and genetically heterogeneous condition. Clinical manifestations range from only mild growth retardation to lethality. Although these are rare, disorder of skeleton are of clinical relevance because of their overall frequency. Skeleton disorder have been subdivided into dysostosis, defined as malformation of bones and groups of bones and osteochondrodysplasia, developmental disorders of chondro-osseous tissue. (1) CT Scan have important role in diagnosis, specially 3D CT has added advantage in identifying bony architecture.

Aim: To evaluate the role of CT imaging in diagnosis of pediatric skeleton disorder, that further helps in management of the disorder. CT imaging has advantage like easily available, fast, affordable compared to MRI. So, in most of skeleton disorder CT is diagnosis of choice, bony abnormality can be better evaluated. 3DCT has good bone architecture differentiation, that helps in further management

Material and Methods: Five case reports of pediatric congenital and developmental skeleton disorder has been discussed.

All the five patients underwent CT scan on Philips brilliance 256 slice machine.

CT imaging findings with 3D Reconstruction have been described.

Case Summary:

Case 1 :Fibrous dysplasia

Case 2 :Developmental dysplasia of hip

Case 3 :Severe kyphoscoliosis

Case 4 :Spinal dysraphism

Case 5 :Osteogenesis imperfecta

Conclusion: CT imaging is most important modality for diagnosis as well as severity of skeleton disorder and that helps in further management of child. Ideal need of ct scan in child, evokes significant stress in parents, even in pediatrician also, but its important to understand that modern machine and CT protocol optimize less radiation to child. Besides that, for certain indications and clinical settings pediatric CT remains preferential over pediatric MRI, e.g., assessment of cortical bone lesions, children with metal rods (e.g., after surgery for scoliosis, limb-lengthening, or osteogenesis imperfecta).

Keywords: pediatric congenital and developmental skeleton disorder; computed tomography.

1. INTRODUCTION

Pediatric congenital and developmental disorder is common nowadays. That particularly occurs due to error in normal development of organ and tissue development. Genetic disorders affecting skeleton comprise a large group of clinically distinct and genetically heterogeneous condition. Clinical manifestations range from only mild growth retardation to lethality. Although these are rare, disorders of skeleton are of clinical relevance because of their overall frequency.

Skeleton disorders have been subdivided into dysostosis, defined as malformation of bones and groups of bones and osteochondrodysplasia, developmental disorders of chondro-osseous tissue. CT Scan have important role in diagnosis, specially 3D CT has added advantage in identifying bony architecture.

Fibrous dysplasia :It is a benign intramedullary fibro-osseous lesion. It is bone anomaly characterized by replacement of normal marrow bone by fibrous tissue that contains a newly formed mineralized product. About 3% patients associated with skin pigmentation and hyper functioning endocrine disorders known as a McCune Albright syndrome. CT imaging, which delineates morphological changes in bone, is modality of choice and superior to x rays.

Spina dysraphism means defective fusion posterior vertebral elements, leading to bifid configuration of spine. It is often considered as a most common CNS malformation. CT imaging has role in diagnosis and severity of defect, according to that further management can be done.

Kyphoscoliosis means increased lateral curvature of spine with Cobb's angle >10 degree, CT has role in diagnosis and in identifying underlying bone abnormality.

Developmental dysplasia of hip refers to abnormal relation of femoral head to acetabulum. CT performed in patients diagnosed with DDH is important in the radiological algorithm because of the advantages as of the axial imaging feature, its capability to investigate the conformity of the femoral head to the acetabular cavity and the ability to measure rotation angles.

Osteogenesis imperfecta is heterogeneous group of congenital, non-sex linked, genetic disorder of collagen type 1 production, CT has role in identify various abnormality.

2. PRESENTATION OF CASES

Five cases of pediatric congenital and developmental skeleton disorder has been discussed.

All the patients underwent CT scan on Philips Brilliance 256 slice CT scan machine at New Civil Hospital, Surat. The informed written consent was taken from the parents/guardians of the patients.

All the patients were subjected to non-contrast CT in axial planes with multiplanar image reconstructions in sagittal and coronal planes wherever necessary and 3D CT.

2.1 CASE I

12 year old female patient presented with chief complains of right sided thigh pain, since 2 years macules in nape of neck since birth.

On plain radiographs, well circumscribed, radiolucent ground glass matrix with cortical thinning is seen.

On CT imaging, Expansile lytic intraosseous bony lesion which shows ground glass internal matrix with some cortical erosion noted involving right iliac bone, acetabulum, sacrum and right femur



Fig. 1. Fibrous dysplasia

2.2 CASE II

8 year old female patient presented with chief complains of left leg pain, shortening of left leg since birth, difficulty in walking since 5 years. On examination, trendelenberg test positive.

On CT imaging, Superior-posterior migration of head of femur noted with head noted in upper outer quadrant on left side. Discontinuity of Shenton's line noted on left side. Head of femur noted above hilgenreiner line. Epiphysis of head of femur on left side appear smaller in size. Shallow acetabulum noted on left side with fibrofatty tissue noted in acetabular fossa.



Fig. 2. Developmental dysplasia of hip

2.3 CASE III

11 year old male patient presented with chief complains of deformity over back region.

On CT imaging, Severe kyphoscoliotic deformity of dorso-lumbar spine with convexity towards right side noted in lower thoracic and upper lumbar spine with crowding of ribs noted. cobb's angle measures more than 40 degree. (severe kyphosis)

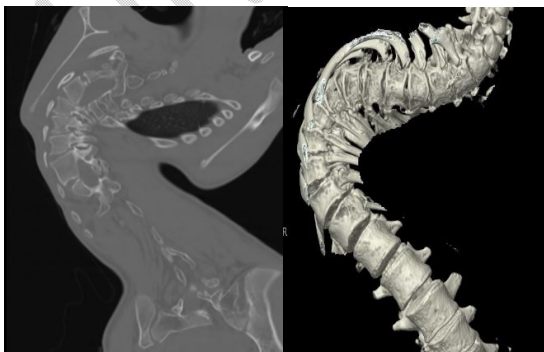


Fig. 3. Severe kyphoscoliosis

2.4 CASE IV

12 year old male presented with chief complain of deformity over lower back region. Cutaneous sinus opening is also there.

On CT Imaging finding, Fusion of Lamina noted between L3-L4 and L4-L5 vertebrae noted on left side. cutaneous.

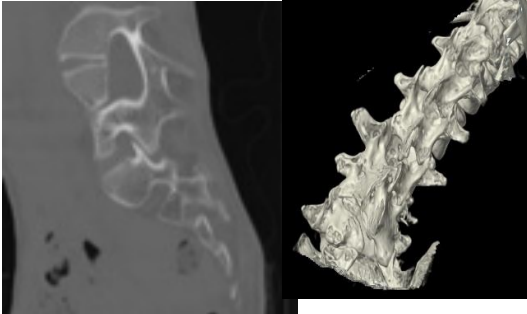


Fig. 4. Spinal dysraphism

2.5 CASE V

11 year old female child presented with chief complain of deformity of lower limb, multiple fractures and difficulty in walking since age of 3 years, multiple operations done to correct deformity and everytimeits develop again and further operation done



Fig. 5. Osteogenesis imperfecta

3. DISCUSSION

Computed tomography (CT) plays an important role in the evaluation of congenital and developmental skeletal disorders in the pediatric patient. With the advent of high-performance 256-section multidetector CT, images can be produced with subsecond gantry rotation times and with submillimeter acquisition, which yields true isotropic high-resolution volume data sets; these features are not attainable with older spiral CT technology. Such capabilities are particularly helpful in the evaluation of pediatric patients by virtually eliminating the need for sedation and minimizing dependence on patient cooperation. The role of three-dimensional (3D) volume imaging in the evaluation of pediatric skeletal disorder continues to evolve, with this technique becoming increasingly important in detection and characterization of lesions as well as in decisions about patient care.

4. CONCLUSION

CT imaging is most important modality for diagnosis as well as severity of skeleton disorder and that helps in further management of child. CT protocol optimize less radiation to child, except certain indications pediatric CT remains preferential over pediatric MRI, e.g., assessment of cortical bone lesions, children with metal rods (e.g., after surgery for scoliosis, limb-lengthening, or osteogenesis imperfecta)

CONSENT

As per international standard or university standard, parental(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

REFERENCES

- 1) **Genetic Disorders of the Skeleton: A Developmental Approach**
Uwe Kornak and Stefan Mundlos
- 2) **Three-dimensional Computerized Tomography and Multiplanar Imaging of Developmental Hip Dysplasia**
MüjdatBankaoğlu
- 3) **Imaging of pediatric bone and growth disorders: Of diagnostic workhorses and new horizons**
Sarah N. Mehany^{1,2} and Janina M. Pat