Ischio-Pubic-Patella Syndrome

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Abstracts: It has been stated that regulation of the development of the iliac bone is different from that of the ischium and pubis. There are well-known clinical syndromes concerned with hypoplasia of ischiopubic bone, such as small patella syndrome, nail-patella syndrome, ischiopubic-patellar hypoplasia, and ischiopubic hypoplasia. A 3 year male child presented with right knee pain since 2 weeks. She sought advice from an orthopedic surgeon and was advised for x-ray right knee. On reviewing the X-ray patella were absent and then further X-rays of left knee , pelvis , spine, elbow and foot were taken on suspicion of ischio pubic patellar dysplasia (IPD) or small patella syndrome. Patient had absent patella on left side and bilateral defective ischio pubic ossification with no spinal, elbow or foot abnormalities.Bilateral inferior ischiopubic hypoplasia along with bilateral patellar aplasia with no spinal, elbow or foot abnormalities probably constitutes a new syndrome. To the best of the author's knowledge, this finding has not been described previously [Marekar A NJIRM 2015; 6(2):115-116]

Key Words: Patella, pubic bone , ischium

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Introduction: In the chaos of malformation syndromes the radiographic detection of ischiopubic patella syndrome, which is an extremely rare anomaly, is particularly helpful in the diagnostic investigation.

The appearance of this anomaly in association with either dysplastic scoliosis with a significant kyphotic element or with a more heterogeneous group of disorders including multiple segmental defects of the spine, rib anomalies and less pronounced facial abnormalities has been described as ischiovertebral dysplasia (IVD) or ischiospinaldysostosis (ISD), respectively. An entity combining abnormalities of the pelvic girdle and aplasia/hypoplasia of the patella was differentiated from the onycho-osteodysplasia (nail-patella syndrome) and has been called ischiopatellar dysplasia (IPD) or the “small patella” syndrome.

Case Report: A 3 year old male child was admitted with complaints of pain in right knee since 2 weeks. Patient was referrred to our department for X-ray of right knee (Figure1). On reviewing the X-ray patella were absent and then further X-rays of left knee , pelvis , spine, elbow and foot were taken on suspicion of ischio pubic patellar dysplasia (IPD) or small patella syndrome. Patient had absent patella on left side (Figure2) and bilateral defective inferior ischio pubic ossification (Figure3) with no spinal, elbow or foot abnormalities.

Figure 1: Radiograph of right knee anterio-posterior and lateral view shows absent patella

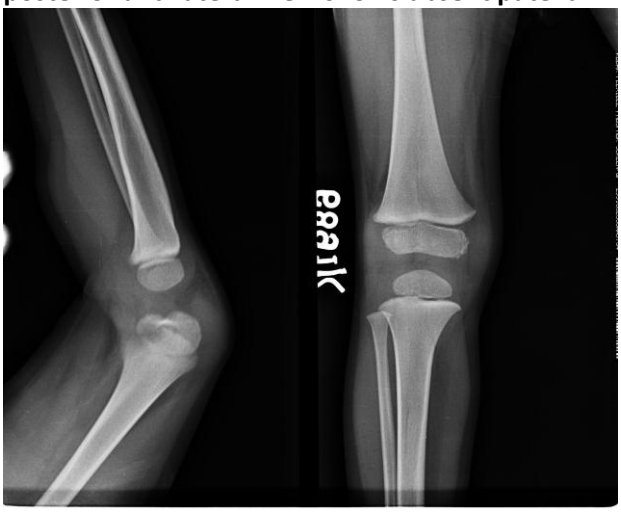


Figure 2: Radiograph of left knee anterio-posterior and lateral view shows absent patella

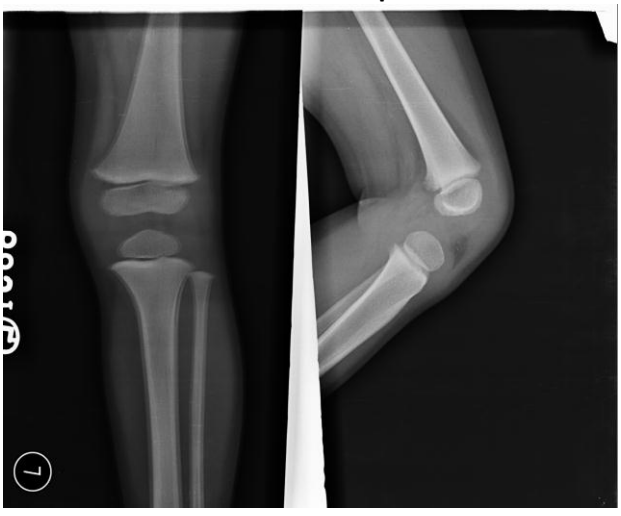
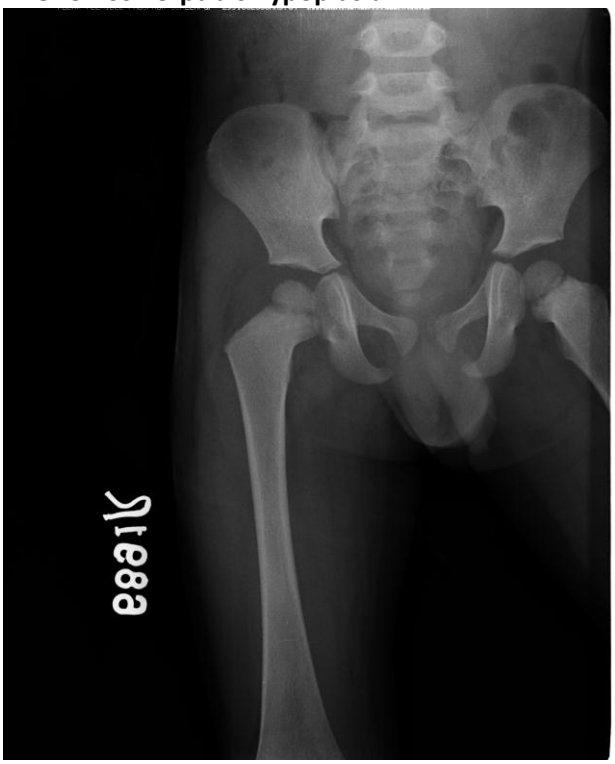


Figure 3: Radiograph of pelvis shows bilateral inferior ischio-pubic hypoplasia.



Discussion: True failure of ossification of the ischiopubic region is uncommon[2]. It has been described in association with several syndromes including acetabular dysplasia, cleidocranial dysplasia, ischiopubic dysplasia (IPD) or the "small patella" syndrome[1,4],IVD,ISD,hypophosphatasia, metatropic dwarfism, campomelic and spondyloepiphysealdysplasia.The clinical and radiological appearances of the patella and pelvis in our patients are consistent with the 'ischio-pubic-patella'syndrome[2,3].Differential diagnosis from the more common and potentially more serious nail–patella syndrome[1] (hereditary osteo-onychodysplasia or Turner–Fong syndrome) was based on the absence of nail dysplasia, elbow deformities or iliac horns.In most reported cases IPD is associated with other malformations such as anomalies of the hips, including CDH, and feet, such as talipesequinovarus.The patient presented exhibited a combination of the syndrome with absent patella that has been previously reported. Although major defects of the ischiopubic bones may occasionally be identified, ischiopubic hypoplasia usually appears as a minor defect of ossification of the ischiopubicsynchondrosis. The appearance of this rare anomaly in young children should always be regarded as a syndromic constituent, and the patient should be carefully evaluated and followed until skeletal maturity.

Conclusion: Hypoplasia of the ischiopubic region together with bilateral aplasia of the patella is an extremely rare anomaly.Lessthat 50 patients have been reported worldwide. Diagnosis is clinical and radiographical. Ischio-pubic-patella syndrome should be recognized and differentiated from other disorders. Early surgical treatment, pain relief therapy and supportive measures should be offered.

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