CURRENT TOPICS IN IMAGING OF JUVENILE RHEUMATOID ARTHRITIS

Preeyacha PACHARN, M.D.[1], Kullanuch CHANCHAIRUJIRA, M.D.[1]
Thomas J.BRYCE, M.D.[2], Thaschawee ARKACHAISRI, M.D.[3]

INTRODUCTION

Juvenile rheumatoid arthritis (JRA) is diagnosed when arthritis is identified in a child aged 16 or younger, lasting 6 weeks or more, in whom other causes of childhood arthritis are excluded. The disease is more commonly found in Caucasian children than African or Asian children.[1] JRA typically demonstrates more rapid progression of disease compared with adult rheumatoid arthritis and can significantly impair bony development in the growing child.

According to the American College of Rheumatology, JRA can be classified into three categories based on number and joints involved, symptoms and the presence of specific antibodies.

(1) The pauciarticular form is the most common form of JRA, involving 4 or less joints.
(2) The poly articular form, in which 5 or more joints are involved.
(3) The systemic form of JRA, or "Still's disease." In this subtype, other organs are sometimes involved. The patients usually develop high fever with rash.

The symptoms of JRA include swollen, stiff, erythematous and painful joints. High fever with rash, lymphadenopathy and fatigue can be seen in systemic JRA. Early diagnosis is crucial. However, there is no single 'gold standard' diagnostic test for JRA. Diagnosis is based on a variety of data including clinical signs and symptoms. There are a large variety of synovial and non-synovial conditions which may mimic JRA[2] Therefore, a careful history and physical examination combined with imaging studies should help to distinguish these conditions from JRA, especially in cases with an unusual clinical presentation. During treatment, imaging plays a key role in the evaluation of disease progression and in monitoring the effect of therapy.

Plain radiography

Plain radiographs have traditionally been the primary imaging modality used to aid the diagnosis. They are the most cost-effective examination in the exclusion of the alternated diagnoses.[3] Early in the disease course, plain radiographs sometime show soft tissue swelling, joint effusions, periosteal new bone formation or accelerated maturation of the epiphysis.[2,4] (fig.1) Soft tissue swelling is usually seen after the first week, resulting from periarticular edema, synovial proliferation and accumulation of joint fluid.[5] However, these findings are nonspecific and may be seen in a variety of inflammatory arthritides.

[1] Departments of Radiology Siriraj Hospital, Mahidol University
[2] Department of Radiology, University of California San Francisco
[3] Pediatrics Siriraj Hospital, Mahidol University

Some findings appear late in the course of disease when substantial bone and joint damage has occurred. Periarticular osteopenia can be seen on plain radiographs when there is at least 30% -50% loss of bone mineralization.[6] Joint space narrowing is indirect evidence of articular cartilage damage and bone erosions are seen after significant cartilage loss has occured. These late changes are usually irreversible. (fig.2)

The lack of specificity of radiographs has resulted in an increase in the use of ultrasound and MRI, which are more sensitive in detecting intra -articular abnormalities.

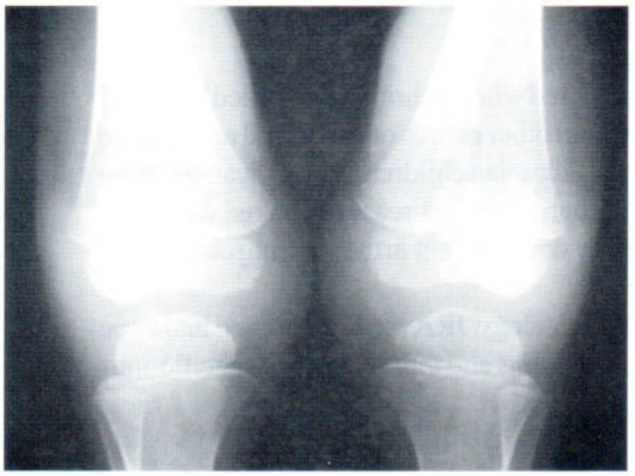

Fig.1A

Fig.1B

Fig.1B

Fig.1 4-year-old girl with history of JRA. Plain radiographs were obtained at the early diagnosis. Plain radiograph of the both knees, AP (A) and lateral (B) view demonstrated marked soft tissue swelling and mild enlargement of epiphysis.

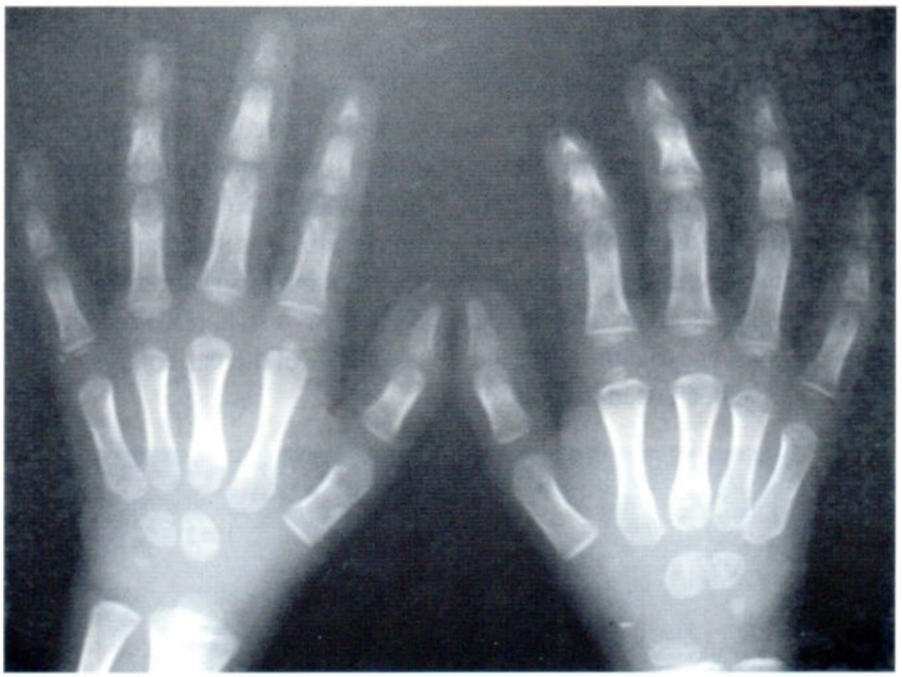

Fig.1 4-year-old girl with history of JRA. Plain radiographs were obtained at the early diagnosis. (C), Plain radiograph of both hands showed soft tissue swelling with no definite bony destruction.

Fig.2 8-year-old girl with JRA Plain radiograph showed diffuse narrowing of carpometacarpal, intercarpals and radiocarpal joint spaces of both hands. There are some erosions of carpal bones.

Ultrasonography

Ultrasound is more sensitive than plain radiographs in detecting cartilaginous erosions, synovial thickening and effusions, but is less sensitive than MRI.[3] A high frequency linear probe (12-15MHz) is usually used. A simple joint effusion is almost always anechoic.(fig.3) An inflamed synovium lining the articular cartilage can be seen as an area of mixed echogenicity.(fig.4A) The Synovial thickness and the amount of joint effusion have been used to monitor disease progress.[7] The vascularity of the synovium can be assessed with Doppler examination. Hyperemia of the pannus is suggestive of the active phase of the disease. Involvement of the articular cartilage can be visualized as thickening in early stages (fig.4B) as well as thinning in later stages. Blurring of the articular cartilage is also noted. Popliteal cysts may be seen in upto 40% of the patients.[5]

Fig.3A

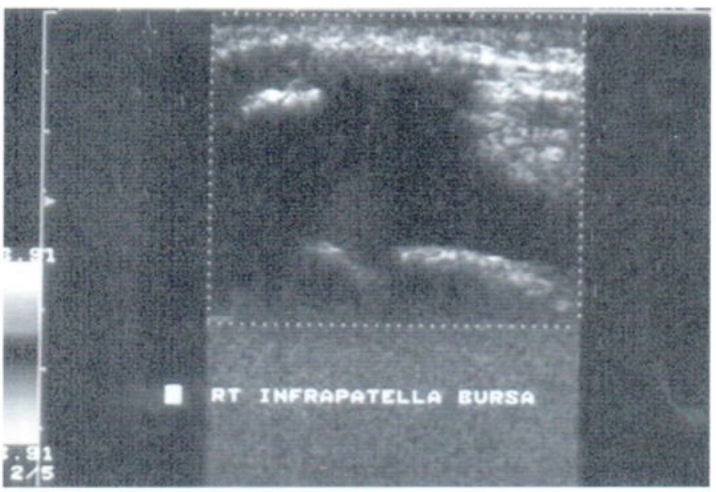

Fig.3B

Fig.3 5-year-old boy with JRA Ultrasound of the right knee at the level of suprapatella bursa (A) and infrapatella bursa (B) demonstrated fluid echogenicity of simple joint effusion.

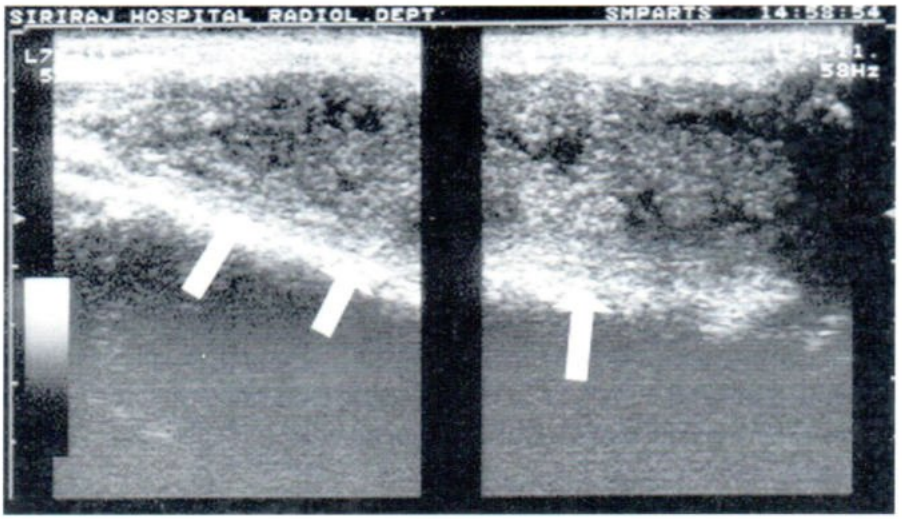

Fig.4A

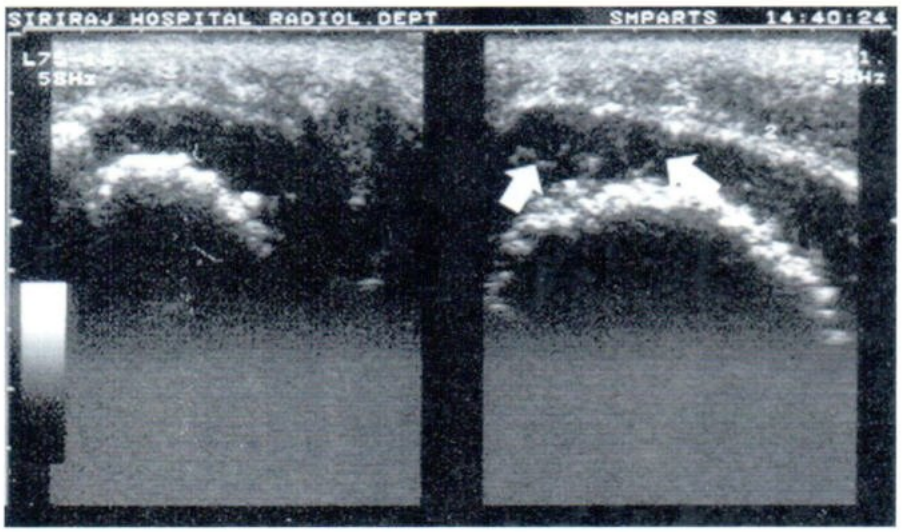

Fig.4B

Fig.4 5-year-old boy with JRA and swelling of both knees (A),Ultrasound of the right knee at the right suprapatella bursa showed distend right suprapatella bursa with mixed echogenic material and thickening of synovial lining.(arrows) (B), Involvement of the articular cartilage in the early stage showed thickening of the cartilage. Hyperechoic strands (arrows) are related to hyperemic vessels.

The advantages of ultrasound are relatively short examination time, noninvasiveness, ease of side to side comparison. However, the technique is operator dependent and requires patient cooperation. It may be difficult to perform in small children. Ultrasound can also be used to aid the aspiration or therapeutic injection of affected joints.

Magnetic resonance imaging (MRI)

MRI is the most sensitive radiologic indicator of disease activity. Because of its excellent soft tissue contrast, synovial hypertrophy, articular cartilage and joint effusions can be clearly demonstrated.[2,8] Peripheral soft tissue can be visualized.

Many pulse sequences are available and different sequences are appropriate for different structures. The selection of examination protocols should be based on the pertinent clinical questions. A small field of view should be used with thin slice thickness.

Bony erosions can be seen using regular T1W and T2W sequences. Hypertrophic synovium is best demonstrated on post Gd-DTPA T1W images with fat suppression. Fibrous, nonvascular synovium is seen as low signal intensity (SI) on both T1W and T2W sequences with no Gd-DTPA enhancement, while inflamed hypervascular synovium shows low SI on T1W sequences and high SI on T2W sequences or T1W sequences with Gd-DTPA enhancement. In inflamed joints, marked enhancement of the thickened nodular or villous synovium is usually identified.[9,10] (fig.5,6) A maximal synovial thickness of 3 mm or more yielded 100% specificity and 77% sensitivity for the diagnosis of synovitis.[10]

Even though synovial enhancement is a non-specific finding, the clinical history and physical examination will be helpful in the exclusion of infection or other joint diseases.

Fig.5A

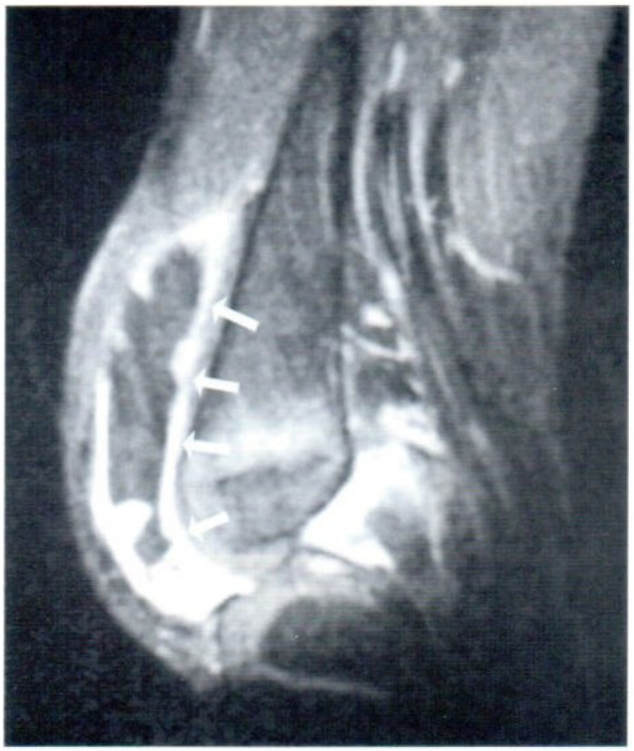

Fig.5B

Fig.5 5-year-old boy with JRA
(A), Sagittal T1weighted image of the right knee showed distension of prepatella bursa with intermediate signal intensity mass like structure, cannot differentiated between thickening synovium and joint fluid. Sagittal T1 weighted after Gd-DTPA enhancement (B) markedly enhancement of thicken synovial tissue is demonstrated. (arrows)

Fig.5C

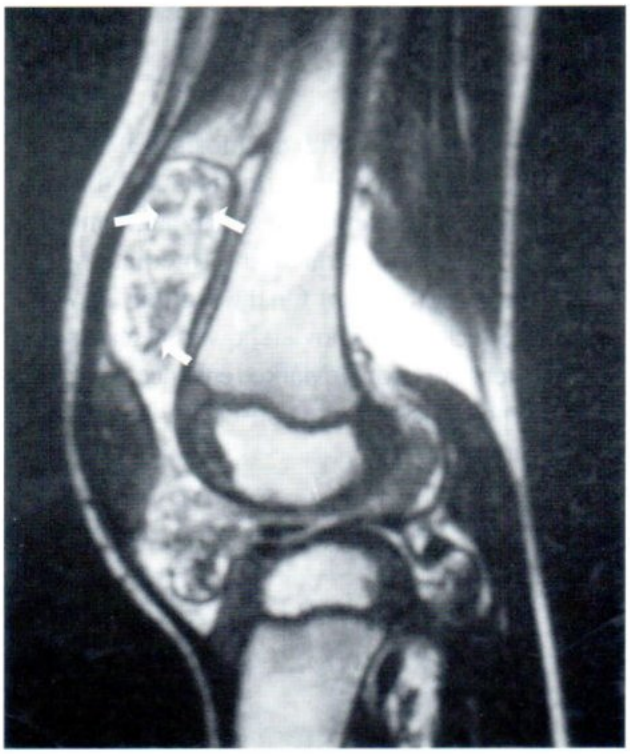

Fig.5D

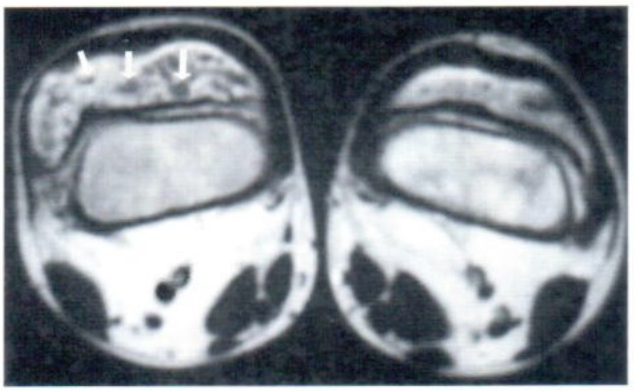

Fig.5E

Fig.5 5-year-old boy with JRA
(C), Coronal T1weight with fat suppression technique and Gd-DTPA enhancement showed vivid enhancement of inflammed synovium in the active phase of disease.(arrows) Sagittal (D) and axial (E) T2weighted images showed synovial membrane with relatively high signal intensity with some joint fluid. Multiple hypo-signal intensity structures, resembling grains of rice inside the joint space is the slough off synovial villi, so called "rice body."(arrows)

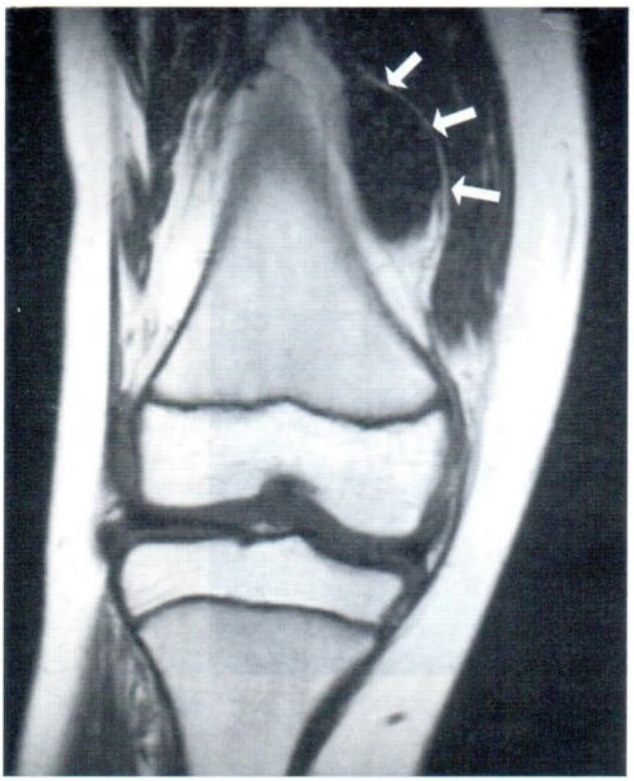

Fig.6A

Fig.6B

Fig.6C

Fig.6 8-year-old boy with diagnosis of JRA Coronal T1weighted image (A) of the knee showed thickening of synovial membrane which demonstrated intermediate signal intensity.(arrows) On the sagittal T2W (B), the thickening synovial showed increase signal intensity.(arrows) Marked enhancement of the synovial is seen after Gd-DTPA injection.(C) (arrows)

Joint effusions show fluid signal intensity, low on T1W and high on T2W sequences(fig.7), which is easily differentiated from inflammed synovium on post Gd-DTPA T1W images.

Articular cartilage is also important because it is one of the earliest structures to demonstrate disease involvement and damage. Cartilage is best demonstrated on FSE T2W or FSE proton density sequences with fat suppression. Cartilage demonstrates high signal intensity on these two sequences. Areas of altered signal intensity, thinning and erosions should be looked for.

A major limitation of MRI is relatively long examination time. The majority of children up to 4-5 years of age require sedation.[11] MRI is also the most costly of the primary imaging modalities for JRA.

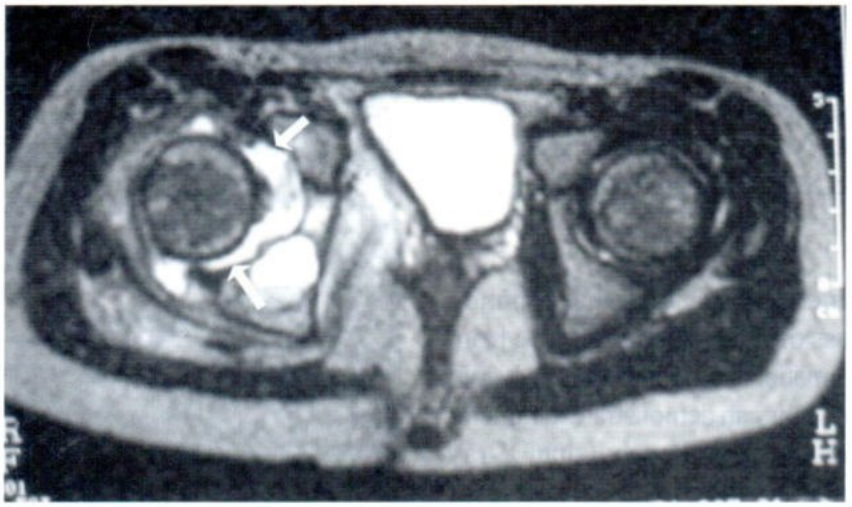

Fig.7A

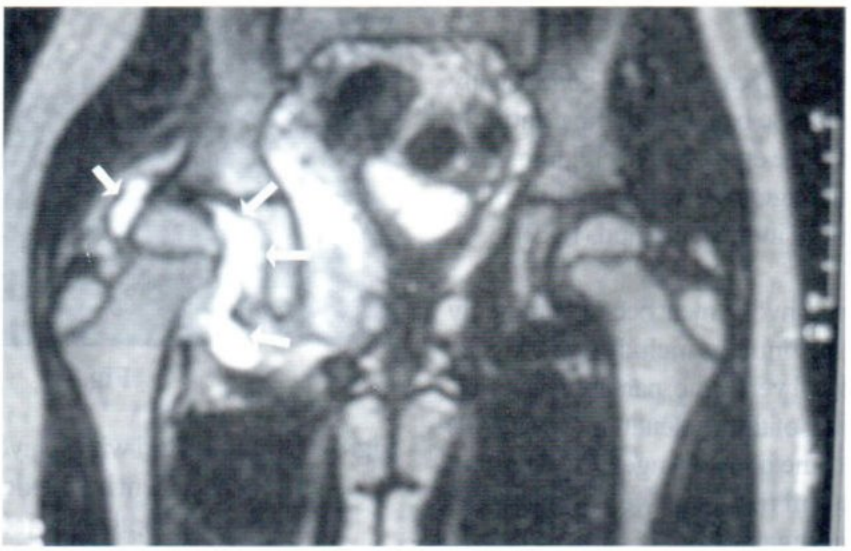

Fig.7B

Fig.7 6-year-old girl with chronic hip pain and diagnosis of JRA Axial (A) and coronal (B) T2weighted images of both hips shows high signal intensity, indicating right hip joint effusion. (arrows)

Complications

Chronic inflammation in JRA results in growth disturbance which can manifest after a few months. (fig.8) Growth disturbance results from hyperemia of the affected joints. Accelerated maturation of the epiphyseal ossification centers, round bones, and sessamoids, and a bulbous appearance of the proximal phalanges can be seen. If the disease occurs before physeal fusion, limb shortening is more likely.[3]

Early treatment will limit growth disturbance. Therefore, there has been an increase in the use of ultrasound and MRI, which are more accurate in the detection of active disease compared with traditional plain radiographs.

Bony demineralization is another common feature of JRA. Loss of bone density may be from hypervascular inflammation, immobilization, or steroid therapy. Plain radiographs can demonstrate demineralization when bone loss is marked, but dual X-ray absorptiometry is more sensitive for lesser degrees of demineralization and is more accurate in monitoring bone mineralization changes.

Erosive changes of cartilage and bone usually appear late in the disease because of the relatively thick cartilage around the epiphyses in children. MRI can detect early cartilage involvement while plain radiographs are only an indirect method.(fig.9)

Cervical spine involvement occurs commonly in polyarticular JRA. Findings include bony fusion, remodelling of cervical vertebrae, paraspinous calcification, and widening of the predental space.(fig.10) Plain film can detect the bony changes, but MRI can be used to evaluate soft tissues as well as the spinal cord and paraspinous ligaments.

Fig.8A

Fig.8B

Fig.8 8-year-old girl with delayed diagnosis of JRA (A), The patient has flexion deformity of both elbows and both knee joints.
(B), Close up image shows marked soft tissue swelling of both hands with fusiform shape of the digits.

Fig.8C

Fig.8 8-year-old girl with delayed diagnosis of JRA (C), Fusion of her left elbow joint is demonstrated.

Fig.9 15-year-old with chronic JRA
Plain radiograph of both hips demonstrated narrowing of both hips joint spaces which reflected the destruction of articular cartilages.

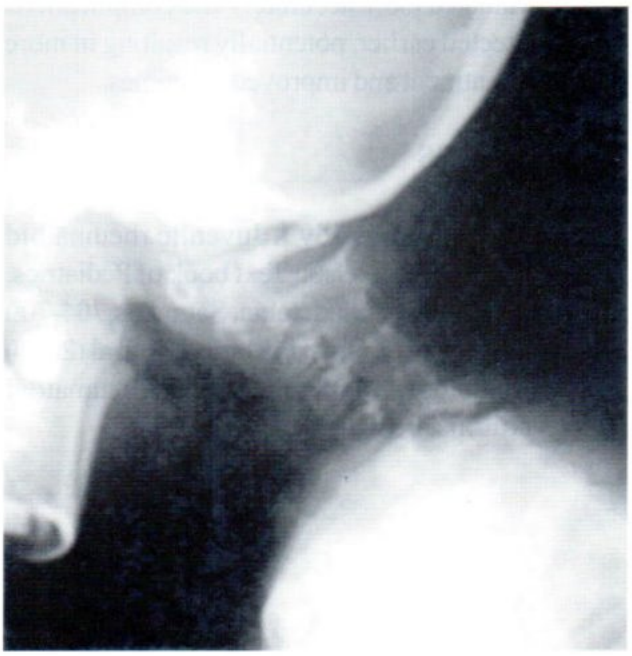

Fig.10 Fusion of cervical spine in a chronic JRA patient.

CONCLUSIONS

JRA is a serious condition which can result in growth disturbance and disability in children. Several imaging modalities are employed for diagnosis and follow up.

Plain film is still quite useful and cost-effective. It may be an appropriate first radiological exam, particularly to exclude alternate diagnoses. However, in children with an atypical presentation, ultrasound or MRI should be considered given that more information about intraarticular structures and processes are provided by these exams, potentially allowing greater specificity in the diagnosis of JRA. For follow up of disease progression, ultrasound and MRI are more sensitive than plain radiographs. Disease activity can be evaluated more accurately and complications can be detected earlier, potentially resulting in more effective treatment and improved outcomes.

REFERENCES

1. Miller M, Cassidy J. Juvenile rheumatoid arthritis. In : Nelson Text book of Pediatrics, 16th edition Philadelphia, Saunders,704-709
2. Wihlborg C, Babyn P, Marilyn R, et al (2001) Radiologic mimics of Juvenile rheumatoid arthritis. Pediatr Radiol 31:315-326
3. Johnson K, Gardner-Medwin J. Childhood arthritis: classification and radiology (2002) Clin Radiol 57:1;47-58
4. Cassidy JT, Martel W. Juvenile rheumatoid arthritis: clinicoradiologic correlations. (1977) Arthritis Rheum:20;207-211
5. Rosendahl K. Imaging of Arthritis in Children: Highlights of Pediatric Radiology : Syllabus 22nd postgraduate courseeuropean society of Pediatric Radiology 1999 May :64-67
6. Resnick D, Niwayama G. Osteoporosis. In: Resnick D.2nd edition. Philadelphia, Saunders, 1988:2022-2085
7. Lamer S, Sebag GH MRI and ultrasound in children with Juvenile chronic arthritis (2000) Eur J Radiol 33:2;85-93
8. Graham T, Blebea J, Gyllys-Morin V, et al (1997) Seminars in Arthritis and Rheumatism 27:3;161-168
9. Johnson K, Wittkop B, Haigh F, et al The early magnatic resonance imaging features of the knee in Juvenile Idiopathic Arthritis (2002) Clin Radiol 57:1;466-471
10. Gylys-Morin V, Graham T, Blebea J, et al (2001) Radiology 220:3;696-706
11. Keengwe I, Hegde S, Dearlove O, et al (1999) Anaesthesia 54(11) :1069-72