

Idiopathic chondrolysis of hip in children: New proposal and implication for radiological staging

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Abstract

Purpose: Our objective was to evaluate the radiological appearances in different stages of idiopathic chondrolysis of hip (ICH) which will be helpful in the early diagnosis and guiding appropriate treatment for this condition to prevent progression of disease. **Materials and Methods:** We evaluated 14 patients of ICH in varying stages: Stage 1 ($n = 9$), Stage 2 ($n = 3$), Stage 3 ($n = 2$). Average age at presentation was 10–11 years. Plain radiograph and magnetic resonance imaging (MRI) was done in all these patients. **Results:** In the current study, we have attempted to stage ICH based on the radiological progression of the disease, where MRI was used as the primary tool. Stage 1 showed a wedge-shaped hyperintensity in T2 weighted (T2W) and hypointensity in T1 weighted (T1W) images involving the middle one-third of the femoral head and it is the earliest and characteristic finding in MRI. Associated findings like joint space narrowing, synovial hypertrophy with joint effusion may also be observed. Stage 2 showed acetabular edema in the affected hip in addition to the above-mentioned findings. Stage 3 showed more extensive involvement of femoral head and acetabulum, with collapse of the femoral head, degenerative changes in hip, early osteoporotic changes, and ultimately loss of joint space. **Conclusion:** Imaging-based staging system proves very useful in the early diagnosis, staging, and assessing the prognosis of ICH.

Key words: Femoral head and acetabulum; idiopathic chondrolysis of hip; radiological staging

Introduction

Chondrolysis of the hip is characterized by rapidly progressive destruction of the articular cartilage of the femoral head and acetabulum with resultant joint space narrowing and restricted movement. Rapid loss of articular cartilage was first described as “acute cartilage necrosis” in the first reported case involving the hip, which was published in 1930 by Waldenstrom.^[1] Chondrolysis involving other major joints like shoulder, knee, ankle, and

elbow have also been reported. Chondrolysis occurs both in children and in adults.^[2]

Slipped capital femoral epiphysis,^[3] severe trauma, prolonged immobilization, infective arthritis, juvenile idiopathic arthritis, and Perthes disease are other known causes of chondrolysis of hip in children. In 1971, Jones^[4,5] reported about cases of chondrolysis of the hip, for which no cause could be identified and hence named as idiopathic chondrolysis of the hip (ICH). Similar cases were also

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reported by Duncan *et al.*^[6] and also by Wenger *et al.*^[7] ICH is a rare pediatric musculoskeletal disorder mainly seen in Africo-Asian adolescents and young children, predominantly in females, as a painful stiff hip of unknown etiology. Synovial fluid aspirate analysis and synovial biopsy can be performed to rule out inflammatory or infectious causes.

Imaging forms the cornerstone for differential diagnosis. Magnetic resonance imaging (MRI) has a high sensitivity and specificity as a diagnostic tool for early identification of ICH.^[8] In the early stages, the destruction appears to be reversible with conservative treatment focusing on pain relief, traction, physiotherapy, and walking with crutches to alleviate weight bearing. Without treatment the disease terminates in spontaneous fusion which needs surgical intervention. Our objective was to stage the disease, based on the radiological progression of appearances, which may be helpful in the early diagnosis to provide best available treatment and assessing the prognosis of this condition.

Materials and Methods

Institutional Ethical Board approval and informed consent for this study was obtained. Our study duration was 5 years starting from 2010 to 2014. A total of 14 cases were studied during this time period. The children included were in the adolescent or preadolescent age group, ranging from 9 to 12 years of age and presented with hip pain and limp. Advanced cases without previous imaging history and diagnosed cases of Perthes disease were excluded from this study. Clinical history was documented. All these children were imaged with plain radiograph of the pelvis and 1.5 T MRI (Magnetom Aera, Siemens, Germany). Computed tomography (CT) scans were performed only for the advanced cases. Retrospective review of the past images of two patients who were being treated as cases of osteomyelitis and arthritis showed the early imaging findings of ICH, which now presented with advanced worsened findings.

The standard protocol for MRI hip was used. Axial, coronal, and sagittal fat suppressed fast spin-echo T2 weighted (T2W) sequence [repetition time/echo time (TR/TE), 3300/31 ms; echo-train length or turbo factor, 8; matrix, 320 × 320; slice thickness, 3 mm; slice gap, 0.7 mm], coronal and axial T1W sequence (TR/TE, 697/17 ms; echo-train length or turbo factor, 2; matrix 512 × 512; slice thickness, 3 mm; slice gap, 0.7 mm), T1W fat suppressed images were obtained in three planes, after intravenous injection of a gadolinium-based contrast agent (0.2 ml/kg). All the images were evaluated by two experienced radiologists. Coronal images were preferred by them for staging because it was easy to define anterior, middle, and posterior third of the femoral head. Synovial thickening and enhancement were diagnosed by

comparing with the contralateral side which were used for staging of the disease.

Plain radiographs were taken for evaluation of each patient's symptoms and those taken for follow-up were reviewed. Joint space width (JSW) was measured at the superolateral, apical, superomedial, and medial aspect and it measures 3–5 mm in normal individuals. Concentric diminution of the articular space to <3 mm or reduction of more than 0.5 mm in the JSW at the narrowest point on plain radiographs on comparison with the contralateral side is considered significant. Two experienced radiologists independently reviewed the routine, follow-up MRI, and plain radiographs and documented the findings. In case of disparity between the observations, agreement was reached by consensus.

The diagnosis of ICH was established in a child with history of limping in the presence of radiographic findings of varying degrees of symmetrical joint space narrowing and MRI finding of wedge-shaped/geographic focal hyperintensity in the central third of the femoral head.

Results

Clinical examination of all the children showed that all of them had hip pain and stiffness in the affected limb. All movements, i.e. flexion, extension, adduction, abduction, and internal/external rotation evoked pain. Full passive range of motion could be achieved with difficulty in the initial stages. Erythema, swelling, or warmth were absent. Muscle atrophy was present in one of the patients with Stage 3 disease. Other joints were spared. All the patient's clinical information and MRI findings were documented as shown in Table 1.

All the cases showed unilateral hip involvement, predominantly involving the right side ($n = 12$). Gender-wise grade analysis shows that there is no significant gender predilection ($P > 0.05$) but 11 out of 14 patients were females. Of the 14 children, nine ($n = 9$) presented with the early features belonging to Stage 1. Three other children ($n = 3$) presented in Stage 2 disease and two children ($n = 2$) presented in Stage 3 ICH with irreversible damage to the hip joint. Almost all the affected children diagnosed were between 9 and 12 years of age, with a peak incidence around 10–11 years of age ($n = 11$).

Early radiographic findings of ICH include varying degrees of symmetrical joint space narrowing (due to loss of articular cartilage), a pelvic tilt toward the side of pain, periarticular osteoporosis, and blurring of the subchondral line.^[9] Protrusio acetabuli is seen in intermediate stage due to softening of the acetabulum and loss of normal growth pattern of triradiate cartilage. Late changes include marginal osteophyte formation, flattening of the femoral head, lateral buttress formation, early closure of the capital

Table 1: Patient characteristics and radiologic features

No	Patient			Clinical features	Radiological findings			Proposed Stage
	Age	Sex	Side		Femur abnormal signal intensity	Joint	Acetabular abnormal signal intensity	
1	11yr	F	R	Hip pain for 6 months	Nil	Space narrowing	Nil	I
2	10yr	F	R	Hip pain& stiffness 6 wk	Geographic Rectangular Mid third, Moderate heterogenous enhancement, Focal enhancement along adjacent cartilage	Space narrowing Mild effusion Synovial thickening	Ill defined Ilium and ischium , Protrusio acetabuli	II
3	9yr	F	R	Hip pain & stiffness for 3 wk	Geographic Rectangular Mid third	Nil		I
4	11yr	F	R	Hip & thigh pain	Geographic Rectangular Mid third	Nil	Nil	I
5	11yr	F	R	Hip pain for 2 wk	Geographic Rectangular Mid third	Nil	Nil	I
6	9 yr	F	R	Hip pain and stiffness 4 wk	Geographic Rectangular Mid third, Moderate heterogenous enhancement, Focal enhancement along adjacent articular cartilage	Joint space narrowing	Nil	I
7	10yr	F	L	Hip pain and stiffness for 4 wk	Geographic Rectangular Mid third	Mild Joint space narrowing	Nil	I
8	12yr	F	R	Hip pain stiffness limping 8 months	Flattening of femoral head, Diffuse increase in femoral head signal intensity osteophytes	Space obliteration Mild effusion Synovial thickening	Extensive ill defined Ilium and ischium, Degenerative changes,	III
9	10yr	F	R	Hip pain and stiffness for 6 wk	Geographic Rectangular Mid third , Moderate heterogenous enhancement, Focal enhancement along adjacent cartilage	Moderate Joint space narrowing Mild effusion Synovial thickening	Nil	I
10	10yr	M	R	Hip pain and stiffness 3wk	Geographic Rectangular Mid third Mild diffuse enhancement	Mild Joint space narrowing Mild effusion Synovial thickening	Nil	I
11	11yr	F	R	Hip pain 10 days	Geographic Rectangular Mid third, Moderate heterogenous enhancement	Mild joint effusion	Nil	I
12	10yr	M	R	Hip pain and stiffness 8 wks	Geographic Rectangular Mid third > 1cm, Moderate heterogenous enhancement	Moderate joint narrowing Mild effusion Synovial thickening	Ill defined Ilium side	II
13	11yr	M	L	Hip pain limp and stiffness 8 wks	Geographic Rectangular Mid third > 1cm Moderate heterogenous enhancement	Moderate Joint space narrowing Mild joint effusion Synovial thickening	Ill defined Ilium and ischium Protrusio acetabuli	II
14	11yr	F	R	Hip pain and stiffness, 6 months	Flattening with more widespread hyperintensity of the femoral head, osteophytes	Fibrous ankylosis, small effusion Synovial thickening	Extensive ill defined Ilium and ischium	III

and trochanteric epiphyseal plates, and fibrous ankylosis [Table 2].^[7,10]

Wedge-shaped or geographic focal hyperintensity in the central third of the femoral head in coronal and sagittal images is the earliest finding in MRI, accompanied by ipsilateral ill-defined adjacent acetabular bone marrow edema,^[8] mild synovial hypertrophy, and little or no joint fluid.^[8] Abnormal signal intensity in the ipsilateral adjacent superomedial acetabulum involves the iliac side of triradiate cartilage in the initial stages and subsequently extending to the ischial side. Degenerative changes are observed as the disease progresses in younger age itself. Articular cartilage thinning is directly depicted in gradient sequences and in postcontrast T1W images, as enhancement of the cartilage adjacent to the femoral head hyperintensity.

Staging

There is some confusion and uncertainty about both the natural history and the results of treatment of ICH. But there is growing recognition that the early diagnosis as well as the staging was important both in the prognosis and starting treatment at an early stage to avoid progression causing permanent damage due to this condition.^[11] A standardized protocol for evaluating and staging ICH by both clinical and radiological imaging is still not yet formulated, hence we attempted to do this using our short case series. Most of the current ambiguity and confusion would be eliminated by this, which would help improve management and prognosis of this condition. Our staging system is mainly based on the sequence of pathological events that occur in this condition. It allows accurate quantification in both early and later stages and avoiding the older invasive diagnostic procedures. After carefully analyzing all the radiographs and MR images, a staging system for ICH was then postulated [Table 3].

Stage 0: Stage 0 is the initial stage of suspicion of ICH, even though the diagnosis cannot be confirmed. Plain radiographs are normal; MRI and other radiological investigations are either normal or nondiagnostic. There is a definite lag between the cartilage loss and the appearance of imaging abnormalities.

Stage 1: In this stage, plain radiographs show normal to variable joint space narrowing, but MRI shows the earliest changes in the form of focal T2 hyperintensity of femoral head corresponding to bone marrow edema. A geometric rectangular/wedge-shaped lesion, hypointense in T1W and hyperintense in T2W images, which is centered

in the middle one-third of the femoral head in coronal and sagittal images and in the anteromedial aspect of middle-third of femoral head in axial images [Figure 1] was recognized. This signal abnormality extended from the articular surface to the proximal physis and measured between 7 and 15 mm in dimensions in Stage 1 and Stage 2 of the disease. After intravenous contrast administration, seven of the nine abnormal proximal femoral geometric foci showed mild to moderate diffuse enhancement, suggesting that it is not a simple edema. As the disease progresses to advanced stage, there is progressive widespread involvement, with ill-defined margins in the head of the femur. There can also be associated synovial thickening and joint effusion in the affected hip. In majority of the cases, the affected hip was in a lower position compared to the opposite hip.

Stage 2: Radiographs showed definite joint space narrowing. MRI showed acetabular edema in the affected hip in addition to joint space narrowing and the wedge-shaped hyperintensity involving middle one-third of the femoral head in T2W images [Figures 2 and 3]. Synovial hypertrophy and joint effusion can also be present. The acetabular inflammation involves the adjacent ilium, triradiate cartilage, and later progressing to involve ischium.

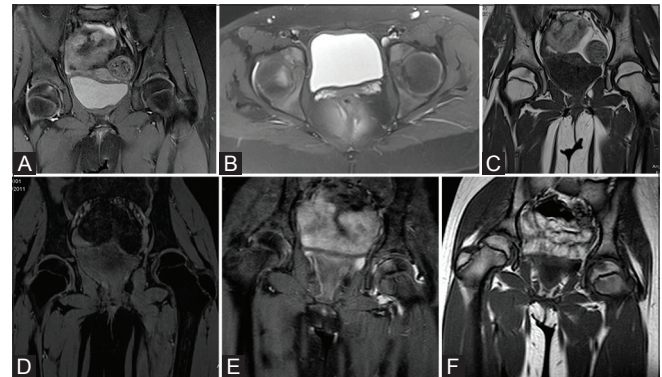


Figure 1 (A-F): 10 year old female child with Stage I of Idiopathic Chondrolysis of Hip (patient 9 in table 1), presented with right sided hip pain and stiffness for 15 days. (A) Coronal fat suppressed T2 weighted image shows a wedge shaped hyperintensity in the middle third of proximal right femoral head epiphysis is characteristic of idiopathic chondrolysis of hip. The acetabulum was normal. (B) Axial T2 weighted image shows a wedge shaped hyperintensity in the medial aspect of right femoral head. Minimal joint effusion is seen. (C) Coronal T1 weighted image shows a wedge shaped hypointensity in the head of right femur. (D) GRE image shows thinning of the articular cartilage overlying the wedge shaped area. (E and F) MRI shows similar findings in the left hip of an 10 years old girl who presented with left hip pain and stiffness

Table 2: Plain x-ray findings in various stages

X-ray findings	Stage I	Total no. Of Stage I cases	%	Stage II	Total no. Of Stage II cases	%	Stage III	Total no. Of Stage III cases	%
Joint space narrowing	5	9	55.5	3	3	100	2	2	100
Protrusio acetabuli	0	9	0	2	3	66.6	0	2	0
Degenerative changes	0	9	0	0	3	0	2	2	100

Stage 3: In this stage, there is widening of the T2 hyperintensity, collapse of the femoral head, extensive acetabular involvement, osteoporotic changes, and loss of joint space due to fibrous ankylosis [Figure 4].

In the final stages of the disease, it is difficult to diagnose this condition due to nonspecific findings which are also found in the end stages of other hip joint diseases. Only retrospective examination of the previous MRI helps in the diagnosis of ICH at this stage.

Table 3: Criteria for radiological staging of Idiopathic Chondrolysis of Hip

Stages	Radiologic findings
STAGE 0	No significant radiological abnormality detected
STAGE I	Variable Radiographic joint space narrowing Wedge shaped focal area of altered signal intensity in the proximal femoral epiphysis which is shown in MRI as focal T2 hyperintensity or T1 hypointensity (Characteristic and Earliest finding in MRI) centered in the middle one third of the femoral head in coronal images ± Synovial hypertrophy and joint effusion
STAGE II	Superomedial Acetabular oedema along the tri-radiate cartilage in the affected hip in addition to joint space narrowing and the wedge shaped hyperintensity involving middle one third of femoral head in T2W images ± Synovial hypertrophy and joint effusion. Protrusio acetabuli may be seen
STAGE III	Widening of the T2 hyperintensity of the proximal femoral epiphysis, collapse of the femoral head, extensive acetabular involvement, osteoporotic changes and degenerative changes with loss of joint space (fibrous ankylosis) ± Overgrowth of femoral head on the neck called "buttress" sign and osteophyte formation

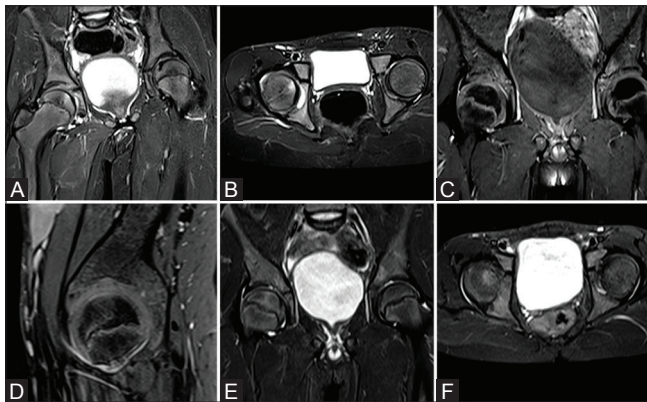


Figure 2 (A-F): (A) A 10 year old female child with Stage II of Idiopathic Chondrolysis of Hip, presented with right loin pain for 8 weeks. (A), Fat suppressed coronal T2 weighted image shows a wedge shaped hyperintensity in the middle third of proximal right femoral epiphysis. Acetabular oedema was seen around the triradiate cartilage involving ilium and ischium. Altered signal intensity noted in the joint space, Pelvic tilting, Synovial thickening and joint effusion is also noted. (B) Axial T2 weighted image shows geographical hyperintensity in the anteromedial femoral head. surrounding muscles appear normal. (C and D) Coronal and Sagittal T1 weighted image with fat suppression after contrast shows mild heterogenous enhancement of the abnormal right femoral head area and junctional zone of the articular cartilage with bone. Visualised left hip shows no abnormal enhancement. (E and F) The followup MRI shows near complete restoration of the joint space and decreased wedge shaped abnormal focus in the femoral head.

One among the children in our study, who was categorized as Stage 3 disease, had undergone MRI 6 months back, which showed Stage 1 findings. The child was being treated as a case of osteomyelitis with antibiotics. The disease progressed from a stage which could be managed conservatively to the stage which had a poor prognosis and thus needed surgical treatment. In one another patient, plain radiograph showed significant joint space narrowing; however, the MRI did not demonstrate any significant abnormality and this patient was earlier treated conservatively for ICH, based on the initial plain radiograph. Absence of MRI abnormality in this patient could possibly represent the radiological improvement in femoral head signal intensity.

We proposed this multimodality imaging-based staging because plain X-ray depicts joint space narrowing, osteoporotic changes, protrusio acetabuli, and degenerative changes, while MRI depicts femoral head and acetabular hyperintensity, joint effusion, synovial thickening, flattening of femoral head and contrast enhancement of the femoral hyperintensity, and articular cartilage. The above radiological findings were analyzed by Fisher's Exact test for their significance [Table 4].

The X-ray findings of protrusio acetabuli and degenerative changes were useful for staging with P value of <0.05 . MRI findings of geographic rectangular femoral head hyperintensity, flattening of femoral head, and acetabular abnormal signal intensity significantly ($P < 0.05$) differentiate

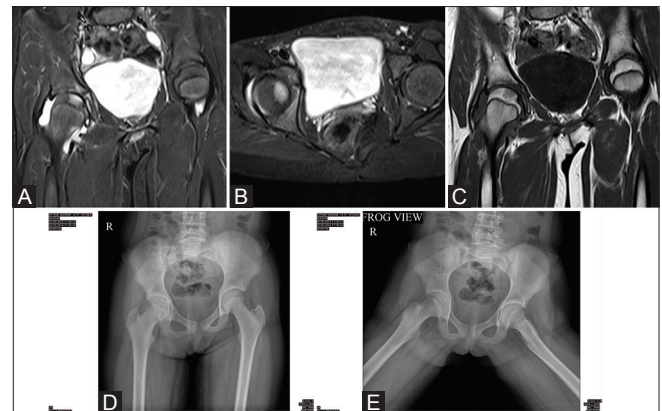


Figure 3 (A-E): 11 year old male child presented with Stage II of Idiopathic Chondrolysis of Hip, who presented with chronic right hip pain. (A) Fat suppressed coronal T2 weighted image shows a wedge shaped hyperintensity in the right femoral head. Acetabular oedema seen around the triradiate cartilage involving ilium and ischium. Joint effusion and synovial thickening are also seen. (B) Fat suppressed axial T2 weighted image shows a wedge shaped hyperintensity in the right femoral head. Acetabular oedema seen around the triradiate cartilage involving ilium and ischium. Joint effusion and synovial thickening are also seen. (C) Coronal T1 weighted image shows wedge shaped hypointensity in the right femoral head. (D and E) Radiographs shows pelvic tilt, diffuse right hip joint space narrowing, periarticular osteopaenia without femoral head deformity and advanced degenerative changes

various stages. Hence each modality is complimentary for staging, but MRI is more sensitive and specific for early diagnosis.

For follow-up imaging both the radiological methods are used. We found that MRI is the only modality to diagnose this condition early enough in Stage 1 to prevent the progression of disease. Hence, if a patient belonging to 9–12 years of age group develops hip pain without an obvious cause and suspected ICH, the initial examination should include MRI with or without plain radiograph of the symptomatic hip.

All our cases underwent treatment, and follow-up imaging was also done [Table 5].

Stage 1 disease with focal femoral head hyperintensity alone is treated with physical therapy, nonweight bearing, and

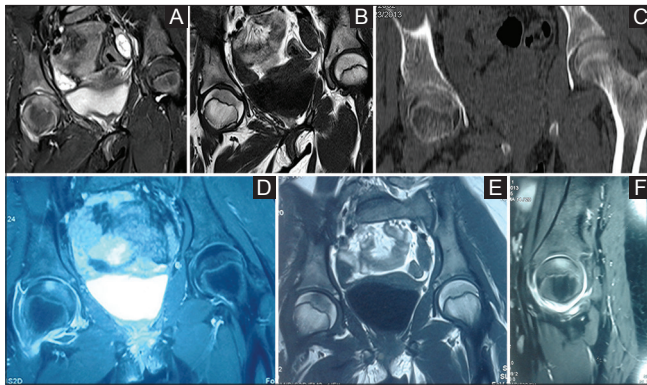


Figure 4 (A-F): A 11 year old female child with Stage III of Idiopathic Chondrolysis of Hip who presented with complaints of limping and right hip pain for a longer duration. Child was earlier treated as some other condition. Patient subsequently underwent total hip replacement. (A) Fat suppressed coronal T2 weighted image shows widening of the abnormal focus in the femoral head with femoral head collapse, acetabular involvement and loss of joint space. (B) Fat suppressed coronal T1 weighted image shows widening of the abnormal focus in the femoral head with femoral head collapse, acetabular involvement and loss of joint space. (C) CT scan shows widening of the femoral head with collapse, acetabular involvement, osteoporotic changes and loss of joint space. Advanced degenerative changes like sclerosis and cyst formation also seen. (D-F) Earlier MRI of the same child which was taken 6 months before shows typical features of Stage I

nonsteroidal antiinflammatory drugs. Soft tissue release and traction were occasionally used to treat contractures. However, many cases resulted in a poor outcome with fibrous ankylosis and loss of hip movement in earlier reports,^[12] probably representing advanced stage. Casting or any other immobilization is contraindicated as the pathologic process involving the articular cartilage may be compounded. Till today, there is no effective therapy to arrest the disease but newer medical therapy aiming at the inhibition of enzymes and other immune mediators of chondrolysis may halt the progress of the disease. Recently, it has been suggested that etanercept, a tumor necrosis factor inhibitor used in the early stages helps in significant improvement.^[13] Conservative treatment can be tried at least for a period of 1 year.

Patients with Stage 2 disease are treated with subtotal circumferential capsulectomy and muscle release, followed by an aggressive rehabilitation program consisting of split Russell's traction, continuous passive motion, and limited weight bearing as reported by Roy and Crawford.^[14] This was supplemented by active and passive range of motion exercises, resulting in resolution of symptoms, improvement in the range of motion, and widening of the joint space, rather than the usual progressive course, but, however, the long-term outcome is poor.^[12] Toward the later part of Stage 2 ICH, arthrodiastasis with external fixation^[15] can be done. Once the condition has progressed to Stage 3, it becomes very difficult to treat and the prognosis is also poor.

Short-term follow-up for 3–4 years in patients with Stage 1 disease show near complete disappearance of the focal femoral head hyperintensity in MRI and the radiograph shows up to 2.5 mm restoration of the joint space in seven of the patients. One patient showed complete disappearance of femoral head edema in MRI, but joint space narrowing still remained. Follow-up of patients showing good clinical improvement in Stage 2 disease revealed no change in imaging. Hence there is no correlation between radiographic signs and clinical recovery.

To date, seven patients in our study have a normal or nearnormal hip joint on radiography, five have moderate to

Table 4: Radiological findings by Plain X-ray & MRI were analysed by Fisher's Exact Test

No	Modality	Findings	Stage I	Stage II	Stage III	Fisher's Exact Test P
1	Radiograph	Joint space narrowing	5/9	3/3	2/2	0.365
2		Protrusion acetabuli	0/9	2/3	0/2	0.043
3		Degenerative changes	0/9	0/3	2/2	0.011
4	MRI	Geographic Rectangular Femoral head hyperintensity without acetabular changes	9/9	0/3	0/2	0.001
5		Hyperintensity in the acetabulum	0/9	3/3	2/2	0.001
6		Hyperintensity in the acetabulum without flattening of femoral head/degenerative changes	0/9	3/3	0/2	0.004
7		Flattening of femoral head + Fibrous ankylosis	0/9	0/3	2/2	0.011
8		Mild joint effusion	3/9	3/3	2/2	0.083
9		Synovial thickening	2/9	2/3	2/2	0.119
10		Fibrous ankylosis	0/9	0/3	1/2	0.143

Table 5: Stage, treatment and outcome in all the cases with idiopathic chondrolysis of hip

Stage	Institution Treatment protocol	Outcome	Follow-up imaging
Stage I (n=9)	Control of synovial inflammation with non-steroidal anti-inflammatory drug/TNF inhibitor, protect weight bearing and maintenance of range of motion	Significant improvement (n=7) No clinical and radiological improvement (n=2)	Complete disappearance of femoral head oedema in MRI in 7 patients
Stage II (n=3)	Partial capsulectomy followed by traction and aggressive rehabilitation, soft tissue release with anti-inflammatory agents	Clinical satisfactory improvement Long-time outcome is poor	No significant radiological improvement in early imaging
Stage III (n=2)	Arthrodiastasis with external fixation Total Hip Replacement	Poor	No significant radiological improvement

severe joint space narrowing of the affected side, and all of the patients are under follow-up. We evaluated our system for reproducibility of assessment, and for the identification of radiologic findings. Plain radiograph and MRI studies of 14 patients were evaluated by two independent radiologists. The resultant data were analyzed by a statistician using inter-rater agreement statistic Kappa with 95% confidence interval for the staging by the two radiologists. A kappa value of 0.20 or less indicates poor, 0.21–0.40 fair, 0.41–0.60 moderate, 0.61–0.80 good, and 0.81–1.00 shows very good agreement. The overall interobserver concordance between radiologists for the staging of ICH was very good (kappa value: 0.901).

Discussion

ICH, first described by Jones in 1971,^[2] is a condition characterized by an extensive loss of articular cartilage of the femoral head and acetabulum, in the absence of known etiologies of chondrolysis like slipped capital femoral epiphysis, trauma, septic arthritis, toxic synovitis, prolonged immobilization, tuberculosis, monoarticular rheumatoid arthritis, Legg-Calves Perthes disease, pigmented villonodular synovitis, leukemia, synovioma, or other neoplasms. It is a condition seen in adolescent and preadolescent children, characterized by progressive loss of articular cartilage of the hip joint due to unknown etiology. The patient, usually an afebrile adolescent female, presents with insidious onset of hip, thigh, or knee pain and limping. Clinical examination in ICH may show a positive Trendelenberg sign. Restriction of the joint motion in all directions can occur in late stages with fixed flexion, abduction, or adduction deformities. The usual causes of hip pain in a child as mentioned above should be ruled out first by a combination of clinical and laboratory examinations. Laboratory findings give negative results for inflammatory parameters, rheumatoid factor, antinuclear antibodies, and culture negative from the joint fluid aspirate.

The true incidence of ICH remains unknown. In general, on comparing with the old case studies, the current study shows no significant difference in the gender predilection, age of onset, and the predominant side of involvement. ICH is more common in girls with a ratio 5 or 6:1. The disease is mostly unilateral, with predominant right-sided involvement. Rarely it can be bilateral.^[16] Right-side predominance can

be due to dominant weight bearing-related trivial trauma. Onset is most frequently around the age of 9–12 years of age in females raising the possibility of hormonal influence and growth spurt, but it can occur in adults also.^[2] The disease is a subacute process developing into full-blown disease within months. It is probably one of the most frequent causes of degenerative arthritis among young women. But indeed, it is not a universally progressive disease as proved by joint space recovery.

The cause of chondrolysis still remains unknown. Theories include autoimmune process^[17-19] alteration in the synovial nutrition to the articular cartilage, mechanical injury, cartilage dysplasia, ischemia, abnormal intracapsular pressure, and inherent abnormal chondrocyte metabolism.^[20] The above-mentioned factors usually cause bilateral involvement, whereas ICH is predominantly unilateral. Our study shows T1 contrast enhancement in the areas of active inflammation within the cartilage with secondary destruction that confirms the ill-defined immunological nature of the disease. But the acetabular involvement and synovial thickening seen in ICH still remains unexplained. Trans synovial extension to adjacent acetabular cartilage is possible. Genetic predisposition to ICH is a possibility, but here also the right-sided involvement cannot be explained.

Histological features of ICH include thickening of the capsule with edematous changes in the capsule and synovium. There are articular surface changes with fibrillation, fragmentation, and progressive loss of cartilage. An infiltration of lymphocytes is seen within the synovium, and there may be degeneration of chondrocyte nuclei and loss of cells in the lacunae.^[17]

Radiographs may show normal hip in the early stages, which later show up reduction in hip joint space. As opposed to ICH, in infections the hip joint space is usually increased. Monoarticular juvenile rheumatoid arthritis (JRA) rarely affects the hip, and in the early stages, typically demonstrates a widened joint space with joint effusion, and an increased ESR and C-reactive protein.^[21] Thus, the radiographic finding of a narrowed hip joint space in patients presenting with hip pain is definitely an important distinction that narrows down the differential diagnosis. The radiographic diagnosis of fractures, neoplasms, slipped

capital femoral epiphysis (SCFE), and Legg-Calve-Perthes disease^[22] are clearly apparent. Plain X-ray cannot accurately differentiate between Stage 1 and Stage 2; however, it helps in picking up the important finding of joint space narrowing^[7] and degenerative changes in the later stages. No criteria are available to classify the severity of joint space narrowing. Isotopic bone scan reveals periarticular increased uptake within the femoral head and acetabulum, which are highly sensitive but not specific.^[23] CT scan is not useful in the early stages and is associated with added risk of ionizing radiation.

In addition to providing early diagnosis, MRI also allows accurate assessment of the course, severity of the disease, helps in staging of the disease process, in the prediction and evaluation of associated complications, in defining the disease, and in the differentiation of ICH from other epiphyseal lesions. MRI depicts the exact extent of the femoral head and acetabular involvement more precisely and also does not expose the pediatric patient to the potentially harmful effects of ionizing radiation. MRI can diagnose and clearly differentiate Stages 1 and 2. Stage 3, usually being end-stage hip disease, is very nonspecific.

Regarding treatment, a large study is required to validate the best treatment options for the corresponding stages. Even though in early stages results are good, long-term follow-up is needed for assessing the final outcome of the patients. Duration for progression from one stage to another, with and without treatment could not be assessed; however, there is a definite halt in the progression of the disease in treated patients. Since there is no clinical staging available till today, no correlation is done between clinical and radiological staging.

From assessing all the Stage 3 cases, we found that failure to diagnose this condition in early stages can lead to progression of the disease from a stage which is reversible by conservative treatment to an irreversible stage causing damage to the hip joint, flexion contracture of the hip, lumbar lordosis causing difficulty even in standing, which all may need surgical treatment and can result from the progression of the disease. In these later stages, the outcome is poor even after surgery.

Radiologic diagnostic criteria

A 9–12-year-old (girls) presenting with unilateral hip pain and the radiograph showing concentric diminution of the joint space to <3 mm with periarticular osteopenia and pelvic tilting without osteophyte formation are diagnostic criteria of ICH. The geometric lesion with a rectangular/wedge-shaped configuration of hypointense T1W and hyperintense T2W signal abnormality centered in the middle one-third of the femoral head in coronal images are the earliest diagnostic feature in MRI.^[24] In the absence of other clinical and biochemical evidence to other known causes of chondrolysis of hip, ICH should be considered as

a specific diagnosis in adolescent patients with any one of the above characteristics radiograph/MRI findings.^[25] CT scan, nuclear scintigraphy, diagnostic arthroscopy, and biopsy are no longer required.

Conclusion

The radiologic staging of ICH based on MRI and plain radiograph helps us to diagnose ICH early in its course so that the treatment can be started early enough to prevent its progression. According to imaging-based classification, MRI is the primary tool because of its accuracy and ability to diagnose at an early stage; however, plain radiograph is included because of its availability and complimentary role. Treatment methods are variable and have no good results in the late stages with or without surgical interventions. The indications for surgery and clinical staging are less clear and there is no well-defined management protocol for these patients. We designed this staging to standardize appropriate criteria for diagnosis, universal terminology to be used by all the specialties and evidence-based algorithm to aid treatment. Radiologists should be made aware of the various radiological findings of ICH, including the different stages of this condition. Awareness, early detection, and treatment of ICH are imperative because they may prevent catastrophic joint destruction. Research on ICH and its treatment based on our imaging-based staging system will improve outcome significantly because of improved imaging and simple reproducible method.

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Conflicts of interest

There are no conflicts of interest.

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