JBR–BTR, 2010, 93: 247-251.

MR IMAGING FINDINGS OF HAEMOPHILIC ARTHROPATHY OF THE ELBOW IN CHILDREN

L. Jans[1,2], M. Ditchfield[1], A. Gomez[1], S. Madhala[1], K. Verstraete[2]

Haemophilic arthropathy of the elbow is a rare cause of elbow pain in children and adolescents. The purpose of this study is to determine the MR appearance of the spectrum of lesions found in haemophilic arthropathy of the elbow at initial MR imaging. It is important to be aware of the early changes in this entity, since early diagnosis and treatment of the disease may prevent progressive joint destruction.

Key-words: Children, skeletal system – Elbow, MR.

Haemophilic arthropathy (HA) of the elbow is an uncommon cause of elbow pain and swelling in children and adolescents.

HA due to recurrent haemarthrosis is the most common musculoskeletal manifestation of haemophilia and one of the most disabling complications of this disease. Children with severe haemophilia suffer from recurrent and acute joint haemorraghe. The therapy consists of factor VIII or IX replacement infusions (treatment on demand). The aim of this therapy is to keep the deficient factor > 1% of its normal value to convert severe haemophilia into a milder form of the disorder. The importance of starting replacement therapy early has been stressed in recent literature (1-3).

The aim of this study is to define the MR imaging features of HA arthropathy of the elbow at initial MR imaging.

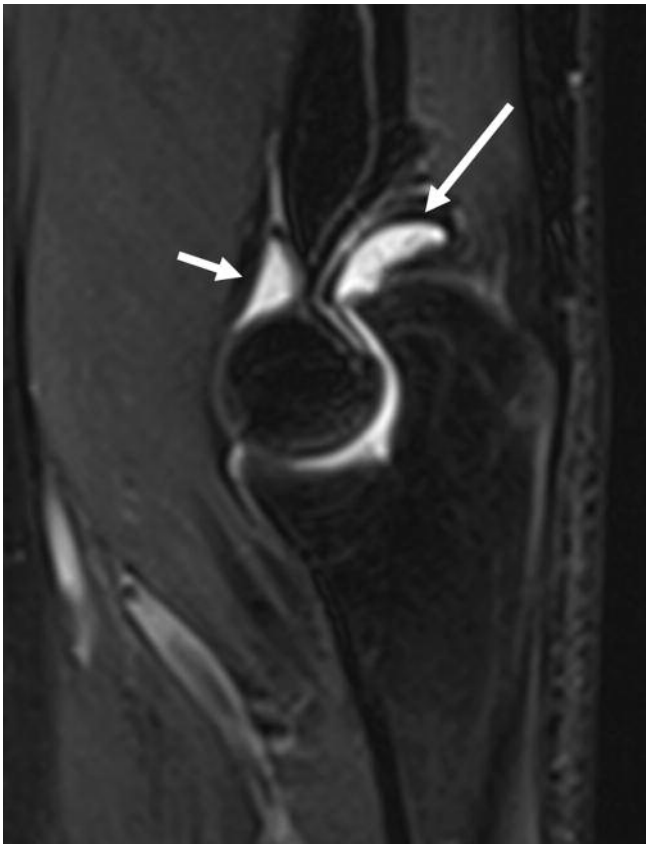

Fig. 1. — Early stage haemophilic arthropathy of the elbow in a 15 year-old boy.
Sagittal T2 PD-weighted MR image. There is a small joint effusion of the elbow predominantly in the coronoid fossa (long arrow) and anterior elbow joint recess (short arrow).

Materials and methods

A retrospective study was performed in a pediatric tertiary care centre. Institutional ethics approval was obtained.

Subjects with documented HA of the elbow were identified using a radiology information system database keyword search of final radiology reports over a 10 year period. Patients with haemophilia presenting with elbow lesions in the setting of acute musculoskeletal trauma were excluded (Fig. 1-5). If the subject had undergone more than one MRI examination, only the first set of MRI images was reviewed. Twelve patients were identified meeting inclusion criteria for the study. The mean age was 12.2 years (range 4.1–16.6 years). All patients had been diagnosed with haemophilia within the first age of life and received factor substitution therapy since.

The images were reviewed by two pediatric radiologists. Both reviewing radiologists were not blinded to the original reported findings but used the report only as a means to identify patients for inclusion into the study.

The presence of joint effusion and synovial hypertrophy was assessed. Synovial haemosiderin deposition was considered present if blooming artefact was evident on gradient

From: 1. Department of Medical Imaging, University of Melbourne, Royal Children's Hospital, Melbourne, Vic, Australia, 2. Department of Radiology and Medical Imaging Ghent University Hospital, Gent, Belgium.
Address for correspondence: Dr L. Jans, M.D., Royal Children's Hospital, Department of Medical Imaging, Flemington Road, Parkville 3052, Melbourne, Vic, Australia.
E-mail: lennart.jans@hotmail.com

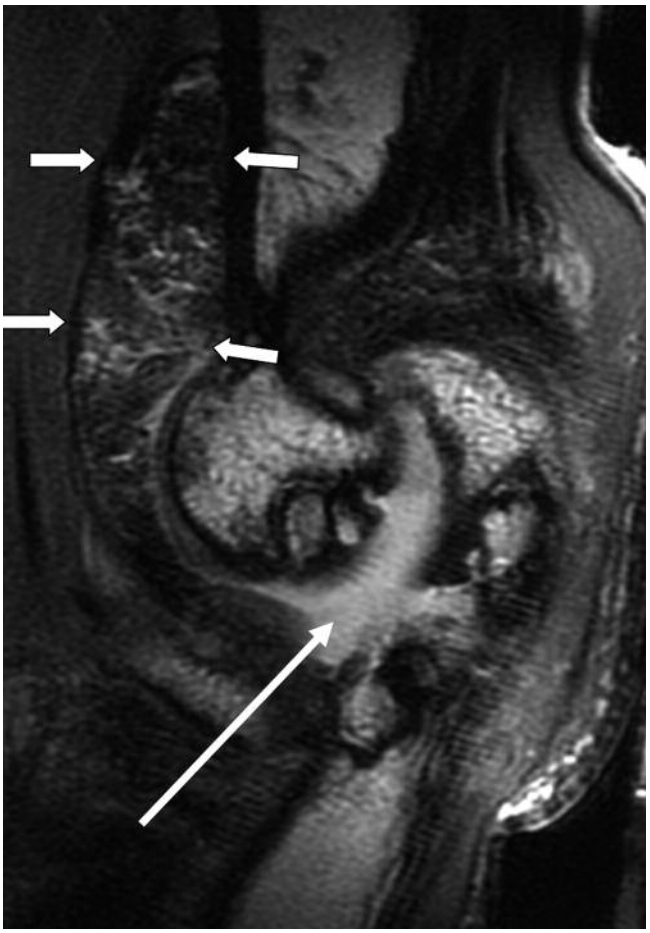

Fig. 2. — Haemophilic arthropathy of the elbow in a 16-year-old boy.

Sagittal PD-weighted MR image demonstrates joint haemarthrosis (small arrows). A large subchondral bone erosion of the olecranon is present (arrow).

echo MR imaging. Joint effusion was graded to be moderate or extensive (extension of all joint spaces). Synovial hypertrophy was graded to be small (< 2 mm), moderate (2-5 mm) or large (> 5 mm). Synovial haemosiderin deposition was determined to be present or absent.

The presence of associated findings including osteochondral lesions, bone erosions, subchondral cysts and presence of loose bodies was analysed. Joint alignment, joint narrowing and epiphyseal overgrowth were assessed.

MR examination

All studies were performed on 1.5 Tesla system (Avanto, Siemens Medical, Erlangen, Germany) with the affected elbow imaged in a small flex extremity four- channel phased circular polarised or array coil (Siemens Medical, Erlangen, Germany). Sequences obtained during MR included sagittal, and coronal oblique fat-saturated T1-

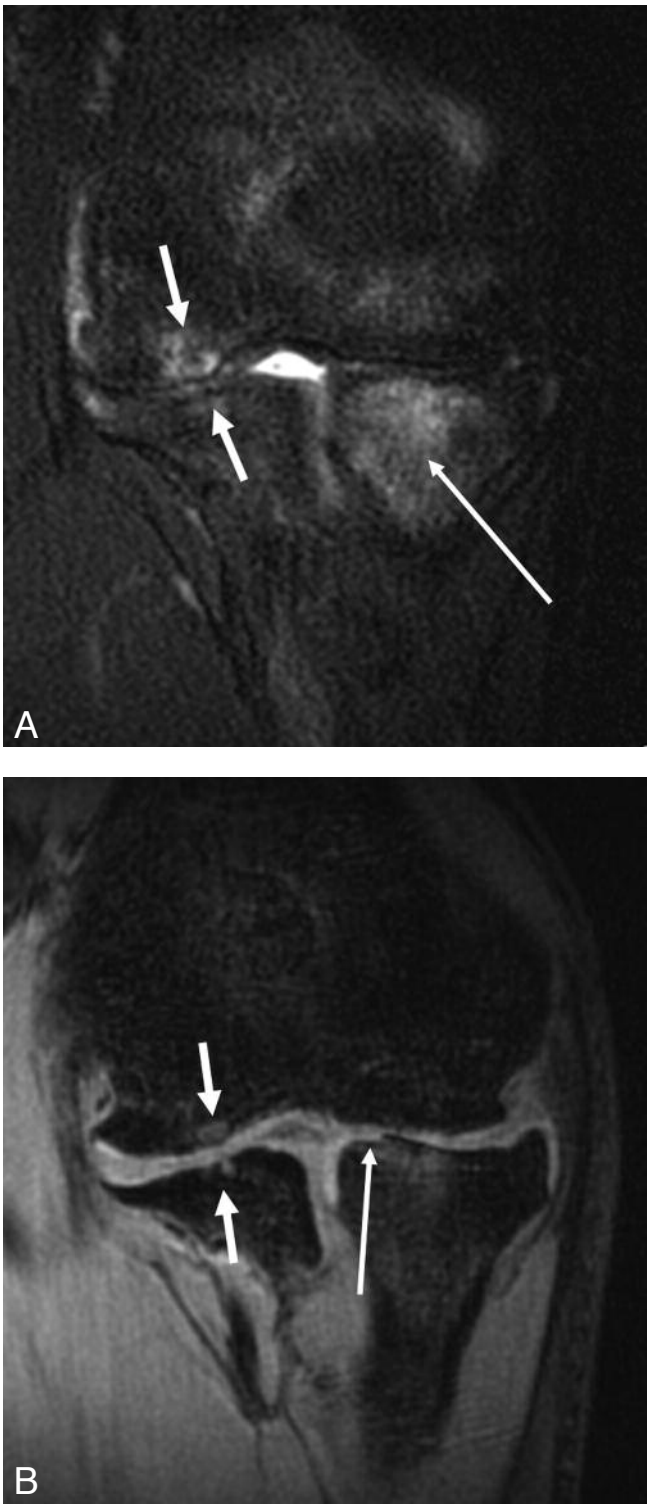

Fig. 3. — Haemophilic arthropathy of the elbow in a 13- year-old boy.

A: Coronal FS T2-weighted MR image. An ill defined area of hyperintense signal abnormality is present on the articular side of the trochlea and ulna (small arrows), in keeping with focal edema. Extensive edema of the radial head is identified (long arrow). B: Coronal FS GRE MR image demonstrates small focal subchondral osseous defects in the articular side of the trochlea and ulna (short arrows), as well as an osteochondral defect of the medial aspect of the radial head (long arrow).

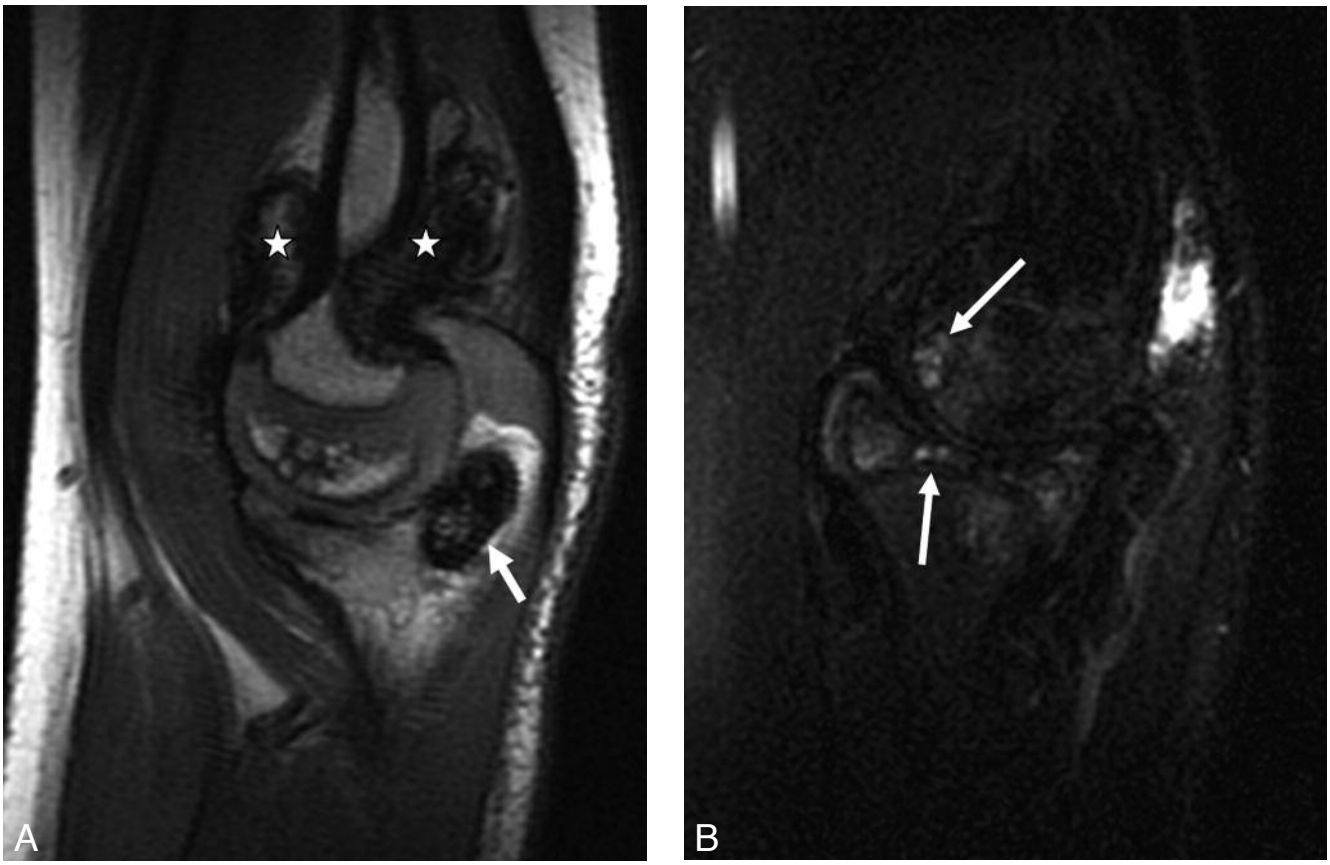

Fig. 4. — Haemophilic arthropathy of the elbow in a 7-year- old boy.
A: Sagittal PD-weighted MR image demonstrates extensive joint haemarthrosis (stars) and subchondral bone erosion in the olecranon (arrow) B: Sagittal FS PD-weighted MR image demonstrates subchondral bone in the capitellum of the humeral physis and in the radial head (arrows).

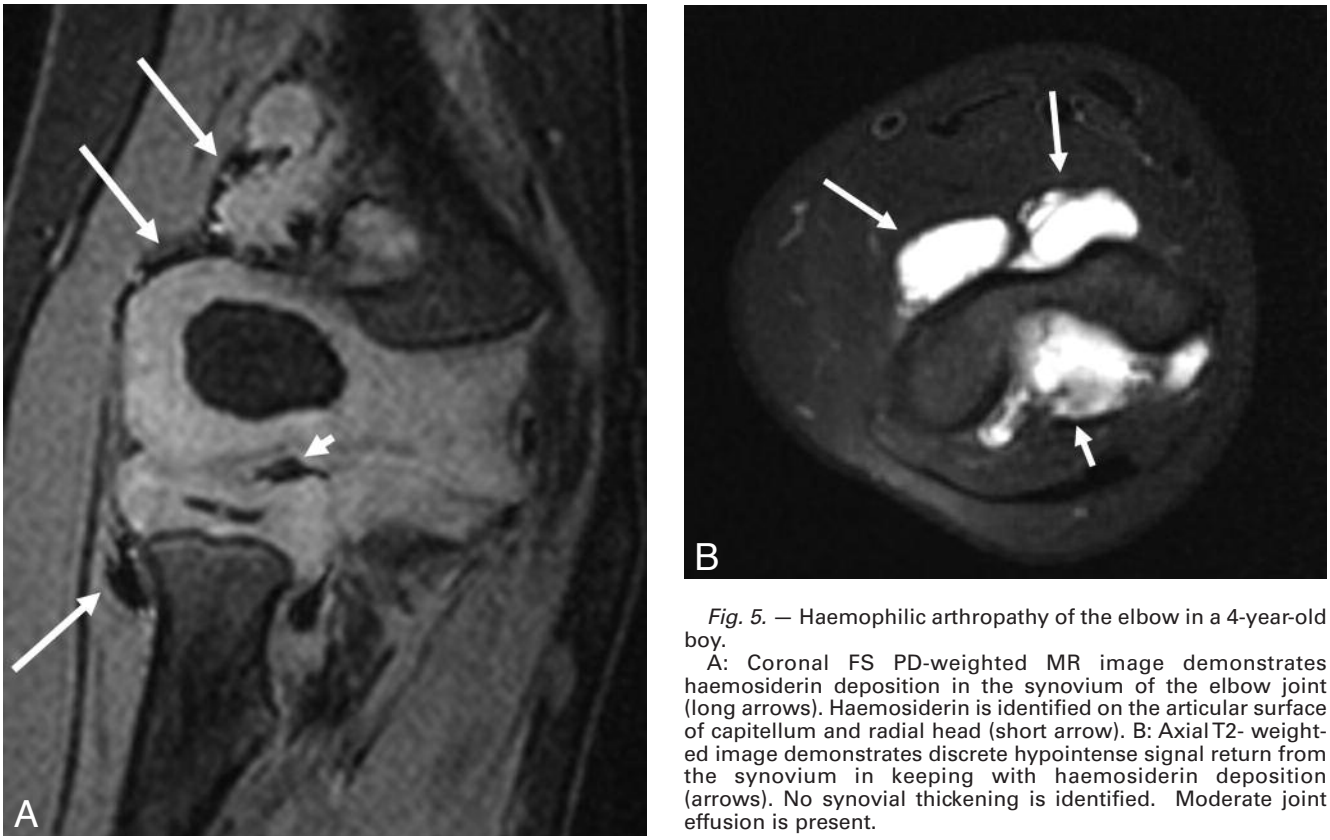

Fig. 5. — Haemophilic arthropathy of the elbow in a 4-year-old boy.
A: Coronal FS PD-weighted MR image demonstrates haemosiderin deposition in the synovium of the elbow joint (long arrows). Haemosiderin is identified on the articular surface of capitellum and radial head (short arrow). B: Axial T2- weighted image demonstrates discrete hypointense signal return from the synovium in keeping with haemosiderin deposition (arrows). No synovial thickening is identified. Moderate joint effusion is present.

Table I. — Petterson radiologic classification of haemophilic arthropathy. Possible joint score: 0-13 points.

Radiologic change	Finding	Score (points)
Osteoporosis	Absent	0
	Present	1
Enlargement of epiphysis	Absent	0
	Present	1
Irregularity of subchondral surface	Absent	0
	Slight	1
	Pronounced	2
Narrowing of joint space	Absent	0
	< 50%	1
	> 50%	2
Subchondral cyst formation	Absent	1
	1 cyst	1
	> 1 cyst	2
Erosions at joint margins	Absent	0
	Present	1
Incongruence between joint margins	Absent	0
	Slight	1
	Pronounced	2
Deformity (angulation and/ or displacement of articulating bones)	Absent	0
	Slight	1
	Pronounced	2

Table II. — The Denver MRI scale classifying the arthropathy in different stages in relation to the most severe finding. The score is progressive, the maximum score is 10.

Score	Stage
0	Normal joint
Effusion/haemarthrosis	
1	Small
2	Moderate
3	Large
Synovial hypertrophy/haemosiderin	
4	Small
5	Moderate
6	Large
Cyst/erosion	
7	One cyst or partial surface erosion
8	More than one cyst or full surface erosion
Cartilage loss	
9	< 50%
10	> 50%

weighted-images; axial and coronal oblique fat-saturated PD (TE = 27 ms) and 3D volumetric spoiled gradient echo sequence.

No direct MR arthrogram or IV contrast administration was performed.

Results

No normal elbow joints were identified. In all patients, MR imaging findings in keeping with HA of the elbow were identified (Fig. 1-5).

Joint effusion was identified in 10 patients (83%). Of these patients, 7 presented with moderate and 3 with large joint effusion.

Synovial haemosiderin deposition was present in 10 patients (83%). Synovial hypertrophy was present in 11 patients (92%). The hypertrophy was small (< 2 mm) in 3 patients, moderate in 2 patients (2-5 mm) and extensive (> 5 mm) in 6 patients.

Chondral lesions were identified in 11 patients (92%). Partial thickness cartilage loss was present in 6 patients; complete cartilage loss was present in 5 patients. Changes of subchondral bone or joint margins were identified in 7 patients (58%). Subchondral cysts were present in 7 patients (58%), all demonstrating multiple cysts in at least one bone of the joint. No patients with articular loose bodies were identified.

Bone marrow edema was present in 8 patients (75%). Focal bone marrow edema in only one bone was identified in 4 patients; extensive multifocal bone marrow edema was identified in 4 patients.

The joint alignment was anatomic in 7 patients (58%) and abnormal in 5 patients (42%). In 3 patients, irregularity of the articular surfaces was identified. Severe misalignment of the elbow joint was demonstrated in 2 patients, with epiphyseal overgrowth in 1 patient.

Discussion

Our study has shown that MRI clearly depicts early changes in haemophiliac arthropathy of the elbow.

In haemophilia, arthropathy predominantly occurs in only a few joints, most notably the knees, ankles and elbows (4). MRI delineates soft tissue, both cartilage and bone and is accurate in the detection of joint effusion and haemosiderin deposition in HA of the elbow. This is why MRI is a precise non- invasive tool for the assessment of early joint changes still undetectable by physical examination or conventional radiography in the haemophilic setting (5).

Early diagnosis of HA of the elbow is mandatory, since it has been stressed that factor substitution therapy from early age decreases the number of rebleeds of the synovial and thus slows down the progressive severity of the arthropathy. Development of late stage disease, with radiosynoviorthesis and ultimately arthroplasty of the elbow, may be prevented (6-7).

Several schemes for the radiological diagnosis, classification and follow- up of HA have been proposed.

The Petterson scoring system of radiological evaluation of haemophilic arthropathy (Table I) is widely recognized (8). This scoring system, however, is based on radiographic findings only. It has been well demonstrated that MR depicts subtle early joint changes in HA that are not depicted by conventional radiography (5).

Several MR scoring systems have been presented that entail two different strategies: the pathology is scored either according to the characteristic stages of development or

Table III. — The European MRI score given in the format A(e:s:h). The A component is the sum of values for changes in following categories: subchondral cysts, irregularity/erosion of subchondral cortex and chondral destruction. The statements are evaluated to weather they are present, for each present feature 1 point is added to the A component. The factors e (effusion), s (synovial hypertrophy) and h (haemosiderin deposition) are evaluated according to a five- point scale. The maximum score is 16 (4:4:4).

Subchondral cysts (part of A)
Present in at least one bone
Present in at least two bones
More than three cysts in at least one bone
More than three cysts in at least two bones
Largest size more than 4 mm in at least one bone
Largest size more than 4 mm in at least two bones
Irregularity/erosion of subchondral cortex (part of A)
Present in at least one bone
Present in at least two bones
Involves more than half of joint surface in at least one bone
Involves more than half of joint surface in at least two bones
Chondral destruction (part of A)
Present in at least one bone
Present in at least two bones
Full-thickness defect in at least one bone
Full-thickness defect in at least two bones
Full-thickness defect involves more than one third of the joint surface in at least one bone
Full-thickness defect involves more than one third of the joint surface in at least two bones
Effusion/Haemarthrosis (e)
Hypertrophic synovial (s)
Haemosiderin (h)
0 Absent
1 Equivocal
2 Small
3 Moderate
4 Large

by summation of specific changes, and thus are respectively 'progressive' and 'additive' methods. The Denver MRI scale shown in Table II represents a progressive scoring system, whereas the European MRI score shown in Table III represents an additive scoring system (9-10). In recent literature combined progressive and additive scales are been developed, resulting in 'compatible scales' (11).

In HA of the elbow, other findings of clinical significance may be present. These features include sprain and tear of ligaments, strain of muscles, presence of soft tissue pseudotumor and late arthropathic degenerative changes. Our study did not focus on these associated findings. However, since these associated findings may be of importance in the clinical setting, awareness of these entities is mandatory.

Gadolinium enhanced MRI may play a role in the differentiation of synovial hypertrophy versus joint fluid. However, contrast administration in the pediatric age group remains a traumatic experience. Given the fact that non enhanced MR imaging provides information of the most common imaging features of HA of the elbow, arthrography was not routinely performed in this study.

Conclusion

The most common findings in haemophiliac arthropathy of the elbow consist of joint effusion, the presence of (osteo)chondral lesions, synovial hypertrophy and synovial haemosiderin deposits. MRI clearly depicts these early changes. Early diagnosis of joint haemorrhage alters the therapeutic management and may prevent progressive joint destruction and surgery.

Acknowledgement

The research was supported by the 2009 research grant of the Royal Belgian Radiology Society (RBRS).

References

1. Soler R., Lopez- Fernandez F., Rodriguez E., Marini M.: Haemophilic arthropathy. A scoring system for magnetic resonance imaging. *Eur Radiol*, 2002, 12: 836-843.
2. Pergantou H., Matsinos G., Papadopoulos A., Platokouki H., Aronis S.: Comparative study of clinical, x-ray and magnetic resonance imaging in evaluation and management of hemophilic arthropathy in children. *Haemophilia,* 2006, 12: 241-247.
3. Utukuri M.M., Goddard N.J.: Haemophilic arthropathy of the elbow. *Haemophilia*, 2005, 11: 565-570.
4. Maclachlan J., Gough-Palmer A., Hargunani R., Farrant J., Holloway B.: Haemophila imaging: a review. *Skeletal Radiol*, 2009, 38: 949-957.
5. Dobon M., Lucia J., Aguilar C., Mayayo E., Roca M., Solano V., Pena S., Giralt M., Ferrandez A.: Value of magnetic resonance imaging for the diagnosis and follow-up of haemophilic arthropathy. *Haemophilia*, 2003, 9: 76-85.
6. Nuss R., Kilcoyne R., Rivard G.; et al.: Late clinical, plain x-ray and magnetic resonance imaging findings in haemophilic joints treated with radiosynoviorthesis. *Haemophilia*, 2000, 6: 658-663.
7. Chapman-Sheath P.J., Giangrande P., Carr A.J.: Arthroplasty of the elbow in haemophilia. *J Bone Joint Surg*, 2003, 85: 1138-1140.
8. Petterson H., Ahlberg A., Nilsson I.M.: A radiological classification of haemophilic arthropathy. *Clin Orthop*, 1980, 149:153-159.
9. Lundin B., Babyn P., Doria A., Kilcoyne R., Ljung R., Miller S., Nuss R., Rivard G.E., Petterson H.: Compatible scales for progressive and additive MRI assessments of haemophilic arthropathy. *Haemophilia*, 2005, 11: 109-205.
10. Funk M.B., Schmidt H., Becker S., Escuriola C., Klarmann D., Klingebiel T., Kreuz W.: Modified magnetic resonance imaging score compared with orthopaedic and radiological scores for the evaluation of haemophilic arthropathy. *Haemophilia*, 2002, 8: 98-103.
11. Doria A.S., Lundin D., Miller S., et al.: Reliability and construct validity of the compatible MRI scoring system for evaluation of elbows in haemophilic children. *Haemophilia*, 2008, 14: 303-314.