Pediatr Radiol (2009) 39:891–898
DOI 10.1007/s00247-009-1233-6

ESPR

Imaging recommendations in paediatric uroradiology

Minutes of the ESPR uroradiology task force session on childhood obstructive uropathy, high-grade fetal hydronephrosis, childhood haematuria, and urolithiasis in childhood. ESPR Annual Congress, Edinburgh, UK, June 2008

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Received: 12 January 2009 / Accepted: 11 February 2009 / Published online: 30 June 2009
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Keywords Paediatric uroradiology · Obstructive uropathy · Childhood haematuria · Childhood urolithiasis

Introduction and rationale

The imaging and procedural recommendations as elaborated by the ESPR Paediatric Uroradiology Taskforce and the ESUR Paediatric Uroradiology Working Group were presented and discussed at the first uroradiology panel held during the ESPR Annual Congress in Barcelona in 2007. These recommendations were published in this journal [1] and have gained wide acceptance throughout Europe and beyond. A few imaging algorithms have been newly proposed by group members, partially completing the existing recommendations, and further important recommendations have been added. The imaging algorithm for prenatally diagnosed hydronephrosis (HN) has been supplemented by a statement on imaging neonates with fetally diagnosed high-grade HN and suspected posterior urethral valves (PUV). Recommendations referring to imaging children with suspected obstructive uropathy, the imaging algorithm for childhood haematuria, and the imaging algorithm for children with suspected urolithiasis have been formulated.

The aim was again to reduce invasive and unnecessary investigations wherever possible without running the risk of missing potentially damaging conditions and increasing patient morbidity. As with the existing algorithms, the new proposals are consensus-based recommendations since little

This article is presented on behalf of the ESPR uroradiology task force and ESUR paediatric working group.

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thorough evidence exists in most of the paediatric queries addressed. The final answer to obstructive uropathy still remains to be found and management of this clinical problem continues to be controversial. How do we define the degree of urinary drainage impairment that puts a kidney at risk of significant damage? After a thorough review of the literature and the existing specific guidelines and recommendations, the pertinent issues were discussed with several paediatricians, paediatric nephrologists, surgeons, urologists, and nuclear medicine specialists. Non-member experts in the field were also consulted.

It is hoped that some standardized imaging procedures for children with these urological conditions will result from these efforts, thus guaranteeing a basic imaging quality at reasonable cost and with a reduction of unnecessary and invasive imaging procedures. The necessary equipment should be made available, not only to major referral centres, but also to less specialized hospitals where children have to undergo diagnostic urological procedures, so that the proposed imaging algorithms might become applicable throughout Europe. Even in the presence of growing economic pressure, proper training in diagnosis, understanding of the findings, and technical knowledge of the procedures should become part of any radiological training programme. Thus, multicentre results should become reliable and comparable.

The second special panel on paediatric uroradiology "Imaging recommendations. Part II" was held at the annual ESPR congress in Edinburgh, UK, in June 2008. The presentation and discussion of the new proposals allowed an open opinion-making process. The results are summarized below.

Imaging algorithm for neonates with prenatally detected high-grade HN, including fetally suspected PUVs

In mild-to-moderate fetal HN initial postnatal US is not emergent and is best postponed until after 1 week of age; these US findings should then dictate additional imaging (Pediatr Radiol 2008; 38:142). High-grade HN, as defined in the adapted SFU-grading system (Pediatr Radiol 2008; 38:139), is frequently associated with severe abnormality, i.e. severe obstructive uropathy and PUV. In some cases, the course may be rapidly deteriorating, e.g. due to acute urinary tract infection (UTI). Therefore prompt diagnosis and treatment are mandatory. Neonates with high-grade fetal HN (particularly if bilateral and with suspected dysplastic parenchyma), with high-grade HN of a single kidney, or those with suspected PUV from fetal US, postnatal US and voiding cystourethrography (VCUG) are emergency procedures, i.e. to be performed during the first day(s) of life. Treatment and further imaging then should be performed according to the severity of the initial findings (Fig. 1). Systematic initial US should always include a thorough assessment of the inner genitalia, particularly in girls, as well as perineal US of the urethra. Using state-of-the-art US techniques, including power Doppler and contrast-enhanced voiding urosonography (ce-VUS) US and VCUG yield adequate information for initial treatment decisions.

The main question in severe HN is whether or not the child needs early drainage and intervention. With the proper answer and adequate urinary drainage instigated, further imaging work-up may be delayed in most cases until physiological immaturity of the neonatal kidney has resolved.

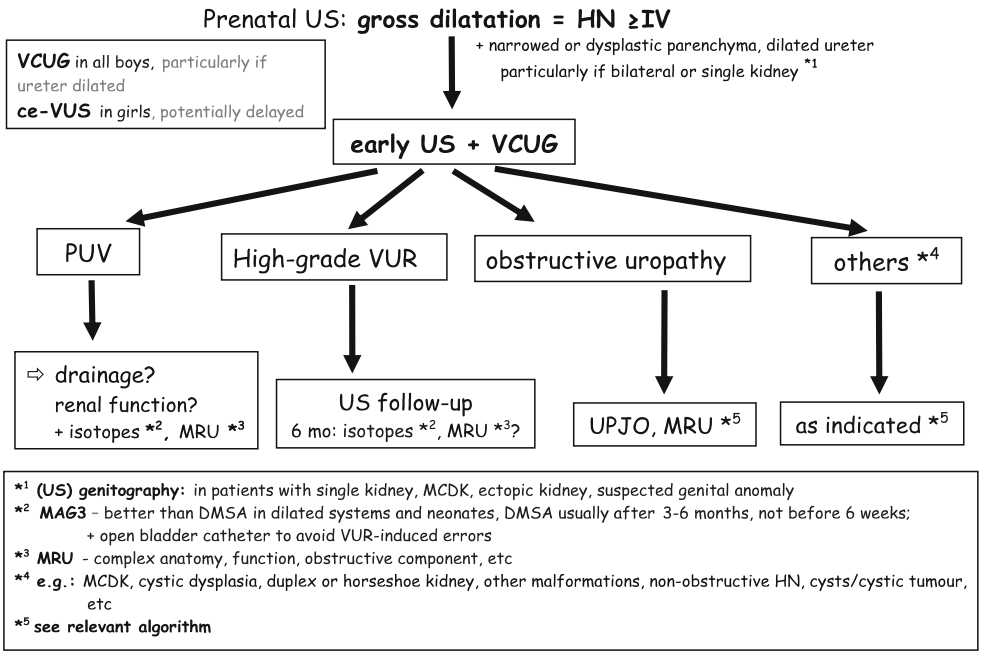

Fig. 1 Imaging algorithm for newborns with fetally diagnosed high-grade HN

This proposal is generally accepted, except for some concerns regarding the liberal use of VCUG, particularly if no dilated ureter or bladder pathology is noted and an underlying ureteropelvic junction (UPJ) obstruction (UPJO) seems more likely. In such cases, assessment of vesicoureteric reflux (VUR) is thought by some to be less urgent, but this depends on local treatment strategies. As with all standard recommendations, some individual adaptations and tailoring to the individual patient's needs and symptoms will be necessary.

Imaging algorithm for children with suspected obstructive uropathy

In children with suspected UPJO or ureterovesical junction (UVJ) obstruction (e.g. primary obstructive megaureter, obstructive ureterocele, etc.) a more sophisticated imaging algorithm is proposed. Initial US will reveal the degree of dilatation of the pelvicalyceal system with or without dilatation of the ureter, presence and degree of ureteral peristalsis (and patency), evidence of urinary tract duplication, or ureterocele, as well as renal parenchymal aspects (thinning, dysplasia, cysts, impaired perfusion, accessory renal vessel that may cause UPJO). Again, further imaging is performed according to the initial US findings (Fig. 2).

It is important to differentiate obstructive and refluxing dilatation. Therefore, evaluation for VUR is highly recommended by VCUG or ce-VUS. In addition, proper assessment of renal function and urinary drainage are indispensible. US on its own is insufficient and may be misleading. Conventional diuretic renography using ^{99m}Tc-MAG3 or (if available) dynamic MR urography (MRU) is recommended. However, even using standardized proto-

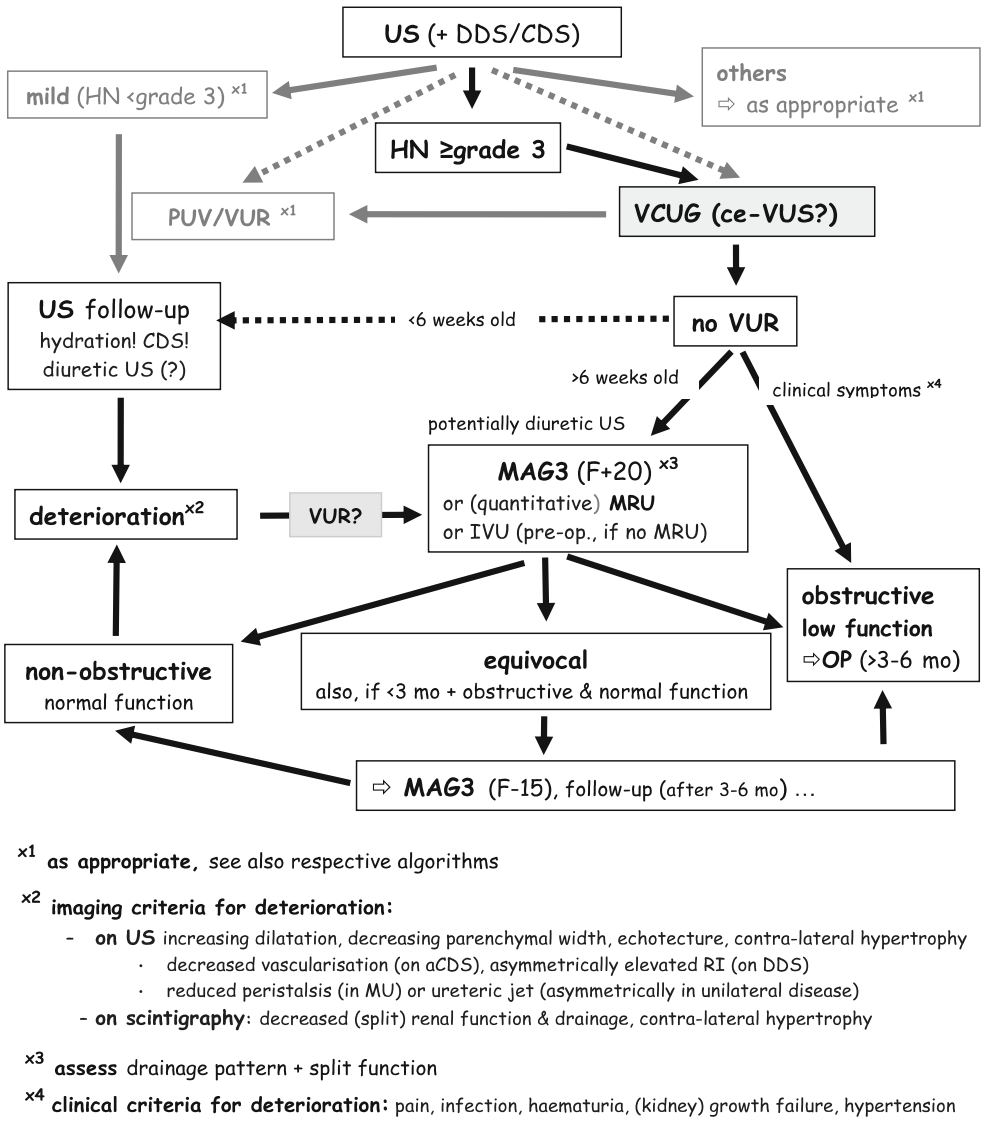

Fig. 2 Imaging algorithm for infants and children with suspected obstructive uropathy

x1 as appropriate, see also respective algorithms

x2 imaging criteria for deterioration:
- on US increasing dilatation, decreasing parenchymal width, echotecture, contra-lateral hypertrophy
  - decreased vascularisation (on aCDS), asymmetrically elevated RI (on DDS)
  - reduced peristalsis (in MU) or ureteric jet (asymmetrically in unilateral disease)
- on scintigraphy: decreased (split) renal function & drainage, contra-lateral hypertrophy

x3 assess drainage pattern + split function

x4 clinical criteria for deterioration: pain, infection, haematuria, (kidney) growth failure, hypertension

cols, including adequate hydration, bladder catheterization and other methodological standards, satisfactory grading of obstruction may be difficult. Particularly during the first months of life, elasticity of the mainly extrarenal collecting system (and, possibly, even the renal parenchyma), and immaturity of renal function may cause confusing results due to the underlying dynamics of these structures.

Because of the unpredictable evolution of the immature nephrourological system and the higher risk of complication, surgery is used with caution during the first months of life and noninvasive close monitoring of further development is advocated. Dedicated ^{99m}Tc-MAG3 scintigraphy using furosemide often helps in differentiating obstruction from equivocal drainage. Even so, there is no overall certainty as to the benefits of early surgical obstruction release for the prevention of acute or future renal functional damage or growth impairment. Furthermore, not enough actual data are available to decide on duration and frequency of follow-up investigations. Features such as increasing pelvicalyceal dilatation with progressive narrowing of renal parenchyma, increased renal parenchymal echogenicity and loss of corticomedullary differentiation, asymmetrical elevation of resistive index, compensatory hypertrophy of the assumed healthy kidney, and deterioration of scintigraphic drainage pattern and split renal function, are considered by many as indicators for surgical intervention. However, even with the proposed imaging algorithm, many uncertainties in the handling of affected individual children may persist.

The aim of this recommendation is an attempt to standardize nephrourological imaging, with the hope to also create comparable data from multiple centres that might yield future evidence for firm decisions and improvement of the various existing imaging algorithm(s). It should increase the awareness that the treatment goal is preservation of renal function and growth potential. The decision for surgical procedures versus noninvasive close observation needs to be based upon combined morphological and functional evaluation. Surgical intervention based simply on the degree of HN might be erroneous and should be undertaken with caution.

Imaging algorithm for children with haematuria

Haematuria in adults may point to a malignant process in the kidney, ureter or bladder and microhaematuria is a common initial symptom of urothelial tumours in older patients. Therefore, aggressive and invasive imaging evaluation by CT urography, supplemented by cystoscopy is indicated in adults. Haematuria in children typically results from a different pathology. Its manifestation should prompt evaluation by US, which may be the only imaging modality required to exclude a mass lesion in the kidneys and bladder.

While urothelial carcinoma in early childhood practically does not exist, gross haematuria may be caused by the rare rhabdomyosarcoma of the bladder (i.e. sarcoma botryoides) or by a renal tumour, typically a nephroblastoma involving the renal collecting system from a breakthrough by the tumour. Nephrolithiasis and urolithiasis, however, are more likely diagnoses in a child with gross haematuria. Most commonly, gross haematuria is caused by some inflammatory process of the bladder (more frequent in girls), or glomerulonephritis. Most important of all, children with the more common manifestation of microscopic haematuria suffer from conditions such as orthostatic or familial haematuria, (glomerulo)nephritis and other nephropathies, UTI, hypercalciuria and urolithiasis, or some systemic diseases; even VUR and obstructive uropathy may manifest with haematuria.

With this wide range of potential abnormalities and the low probability of a malignancy, imaging of a child's urogenital tract does not require the same aggressiveness that is indicated in adults. A thorough clinical evaluation including a detailed family history and laboratory assessment of urine and serum parameters should precede any imaging requests; erythrocyte morphology is often helpful. In general, US examination will reveal the diagnosis and/or exclude a mass lesion. Depending on the findings, additional imaging work-up may then be initiated according to the respective imaging algorithm. An exception necessitating additional imaging procedures in spite of an unremarkable initial US examination is the strong clinical suspicion of a more serious abnormality not correlating with the apparently innocuous US findings (Fig. 3). Note that the main message of this recommendation is to reduce invasiveness and promote the use of thorough US as the primary and in most cases only diagnostic imaging procedure. It is not possible to list all the secondary imaging steps in this vast variety of conditions that may be encountered in childhood haematuria.

Imaging algorithm for children with suspected urolithiasis

Urolithiasis is less common in children than in adults. However, endemic factors combined with nutritional habits render childhood urolithiasis more frequent in some areas. Adult standard imaging protocols cannot be indiscriminately applied to children because of their increased risk from radiation exposure. Other aspects also need to be considered: small and poorly calcified stones are frequent in children (e.g. cystinuria, infectious stones); a child's small ureter with little surrounding fat reduces the diagnostic

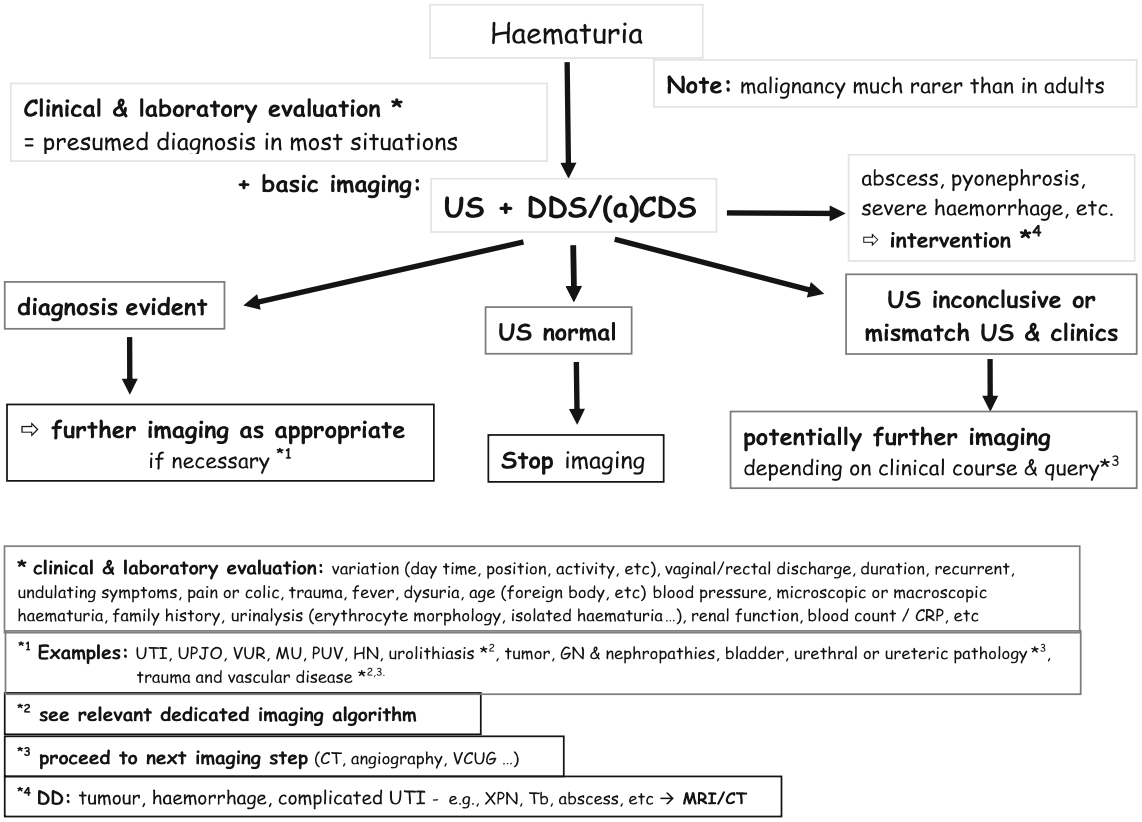

Fig. 3 Imaging algorithm for infants and children with haematuria

potential of CT, particularly when using an unenhanced low-dose technique. The clinical manifestation of urolithiasis may be misleading in infants and small children who typically present with nonspecific abdominal pain, UTI, vomiting, or just haematuria.

On the other hand, US with its entire spectrum of up-to-date techniques is the ideal primary imaging modality in children. It allows reliable demonstration of the kidneys and urinary tract in most cases and may depict indirect signs of abnormality, thus indicating supplementary tailored imaging. Most stones are found in the pelvicalyceal system or in the proximal and/or the distal ureter, i.e. close to or at the UPJ or UVJ, respectively. In children, these areas may be well visualized by US with adequate hydration and urine volume in the bladder. Occasionally, even small concretions are detected that might have been missed by intravenous urography (IVU) or low-dose CT. US criteria such as echogenic foci producing shadows, dilatation of the ureter and pelvicalyceal system, and increased renal echogenicity and size are more conspicuous for diagnosis in children than in adults. The twinkling artefact on colour Doppler US may enhance the suspicion or diagnosis of urinary calculi. Duplex Doppler US may demonstrate unilateral increase of resistive index in acute severe ipsilateral obstruction, thus enhancing the diagnostic potential of US in children.

A plain abdominal radiograph (kidneys/ureters/bladder, KUB) may be necessary for proper stone localization prior to lithotripsy or as a baseline study for follow-up evaluation in selected cases. IVU is still used by some in the diagnostic imaging of paediatric urolithiasis, especially if CT is not available or access to it is limited. In such cases IVU should be performed with restriction, i.e. tailored to the specific question to supplement the information already available from the preceding US examination. IVU limited to three or four views, including the KUB, and with adequate coning will typically be sufficient for diagnosis and will result in a lower radiation dose than traditional standard IVU protocols.

There are very limited data available on low-dose CT in children for the diagnosis of urolithiasis and a lack of information on its diagnostic accuracy for small and poorly calcified stones as frequently encountered in children. It is not known whether the diagnostic accuracy of low-dose CT in adults can be reproduced in children. Thus, we are reluctant to promote a general and indiscriminate use of unenhanced low-dose CT for the diagnosis of urolithiasis in children. Also, emerging reports on the successful use of MRI and MRU for the diagnosis of urinary stones need to be considered. At present, CT may be used as a complementary imaging modality in cases with nondiagnostic or equivocal US findings that do not correlate with the clinical findings,

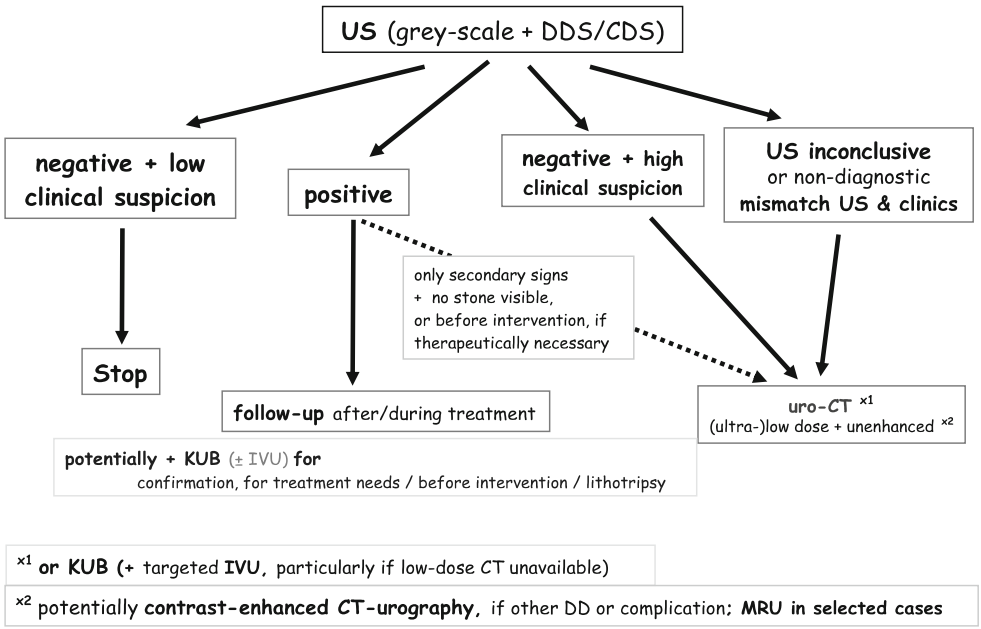

Fig. 4 Imaging algorithm for infants and children with suspected urolithiasis

or if proper sonographic stone localization is not possible, as well as for assessment of complications and for differential diagnosis in a complex case (Fig. 4).

ESPR statement on nephrogenic systemic fibrosis in children

Nephrogenic systemic fibrosis (NSF, or nephrogenic fibrosing dermopathy) is a recently defined disease with a potentially deleterious outcome and not yet completely clarified aetiology. A common factor in many patients is underlying kidney disease (with renal insufficiency, often on dialysis). Another commonly observed factor is repeated gadolinium administration, although there are patients suffering from NSF without previous known gadolinium exposure. Additional independent risk factors are metabolic acidosis and inflammatory and postoperative conditions. Although there are only a few reports describing children with NSF only partly linked with gadolinium exposure, the paediatric radiology community is urged to consider NSF when performing MRI, particularly also considering the potential, yet unknown risk from gadolinium deposition in the bone marrow. In order to properly address these potential risks of gadolinium administration and considering the current discourse, some precautions also seem necessary in children (Fig. 5):

- Reconsider the need of MRI. Sophisticated US can probably solve a number of problems, answering the therapeutically relevant questions, thus making MRI unnecessary.
- There are situations where contrast-enhanced MRI should be reconsidered, and unenhanced MR scans using new techniques can answer the query.
- Performing CT is associated with a radiation burden and contrast-induced nephropathy, and needs to be considered very carefully as alternative imaging.
- Precautions have to be taken to identify patients at increased risk of NSF, i.e. patients with renal disease or after liver transplantation. One may rely on history and clinical data, or ask for a recent blood sample to prove normal creatinine and estimate glomerular filtration rate (GFR).
- In all patients with renal disease or impaired renal function, as well as in patients with inflammatory conditions and acidosis, GFR measurements or estimates should be performed. If below 30 ml/min the indication for contrast-enhanced MRI should be reconsidered and discussed on an individual basis in close collaboration with the attending nephrologist. If (esti-

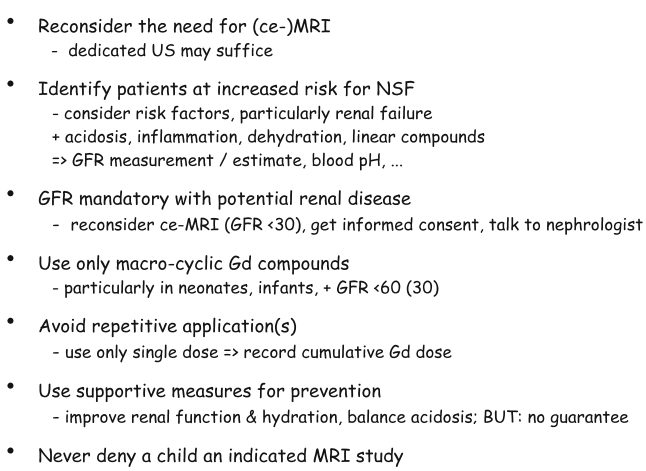

Fig. 5 Gadolinium and NSF in children: what to consider for paediatric MR imaging

mated) GFR is between 30 and 60 ml/min, gadolinium should be administered with caution. Patients and their parents have to be informed about the potential risk and informed consent should be obtained.
- Particularly in patients at increased risk of NSF and in infants, only more stable macrocyclic gadolinium compounds should be used, as they are presently considered to be associated with a lower risk.
- As repeated applications leading to a high cumulative systemic gadolinium dose appear to be a risk factor, reduction of repeat investigations and use of single-dose techniques should be promoted. The cumulative gadolinium dose received by a patient should be recorded and noted, and a thorough follow-up should be established.
- Supportive measures for preventing NSF are balancing acidosis, hydration, and improvement of renal function prior to administration. However, all these measures, even dialysis, do not guarantee full protection.

However, despite the above comments one should never deny any child a well-indicated MRI study that offers therapeutically or prognostically essential information.

Conclusion

The presented recommendations for the various imaging algorithms have been modified according to comments and discussions during the ESPR workgroup session in Edinburgh. The use of VCUG remains controversial, although in many places it remains a standard procedure in the assessment of moderate or severe HN and in infants with an upper UTI or any 'significant' urinary tract malformation. Conversely, some advocate completely dropping the VCUG from routine imaging work-up because of its need of catheterization and radiation, and only using it in selected cases and in special referral centres where high performance and technical standards are guaranteed. No ultimate recommendation can be made at this time based on scientific data. The majority of participants therefore agreed to continue using VCUG (or ceVUS) as part of the primary imaging protocol in all infants with high-grade HN in order to avoid any risk of later renal damage.

Further reading

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