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ORIGINAL ARTICLE

Is renal scintigraphy really a necessity in the routine diagnosis of congenital solitary kidney?

Jera Grabnar[1] · **Rina R. Rus**[2]

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Abstract

Background For a definitive diagnosis of congenital solitary kidney, renal scintigraphy is suggested as being the gold standard of ruling out ectopic functioning renal tissue, possibly missed by ultrasound. The aim of our study was to test ultrasonography precision in comparison with renal scintigraphy on a larger cohort of congenital solitary kidneys.

Methods We performed a retrospective unicenter study of children with congenital solitary kidney with no contralateral tissue, who were treated in the period from 1980 to 2017. The findings in children who underwent both abdominopelvic ultrasound and nuclear renal scintigraphy were compared and the accuracy of ultrasound was assessed.

Results 99 children met the inclusion criteria of congenital solitary kidney confirmed with abdominopelvic ultrasound and nuclear renal scintigraphy. The children were predominantly male (61.6%), and the congenital solitary kidney was largely right-sided (55.5%). In 97 cases (98%), ultrasound correctly predicted the absence of functional renal tissue on one side in the renal fossa or in an ectopic location (pelvis or ipsilateral side). The calculated accuracy of abdominopelvic ultrasound in diagnosing congenital solitary kidney was therefore 98%.

Conclusions Our findings confirm that abdominopelvic ultrasound alone is accurate enough to diagnose congenital solitary kidney. It gives enough information for consideration if further radiological evaluation is still needed.

Keywords Children · Congenital kidney abnormality · Scintigraphy · Solitary kidney · Ultrasonography

Introduction

Congenital solitary kidney (CSK) is the anatomic or functional absence of one kidney from birth, and develops as the result of abnormal or incomplete renal development in utero, leading to a non-functioning kidney as in multicystic dysplastic kidney (MCDK) and renal aplasia, or due to unilateral renal agenesis (URA) [1]. Aplastic/(hypo)dysplastic kidneys or MCDK can spontaneously involute prenatally or in the first years after birth. Spontaneous involution of (hypo)dysplastic kidney is probably the reason that URA incidence data vary according to studies, and is probably lower than previously considered [1–4]. According to Westland et al. [1], the worldwide incidence of URA is around 1:2000 births. CSK can also be associated with other congenital anomalies of the kidney and urinary tract (CAKUT), of which vesicoureteral reflux (VUR) is the most common and is identified in one quarter of URA patients [1]. Additionally, extra-renal anomalies (cardiac, genital or gastrointestinal) were identified in one third of patients with URA [1].

To confirm the diagnosis of URA, dimercaptosuccinic acid (DMSA) [5] or dimercaptoacetyltriglycine (MAG3) renal scintigraphy are recommended and widely used [6]. They are aimed at ruling out ectopic functioning of the renal tissue that was possibly missed by ultrasound (US), and/or evaluating the function of the solitary kidney. In both methods the child is exposed to ionizing radiation, both are time and money consuming, and often require sedation, particularly in young children [7]. On the other hand, renal US is a fast, relatively cheap, safe and non-invasive method for screening urinary anomalies [4]. However, the accuracy of US is user dependent and renal ectopy can be missed or misinterpreted [8, 9]. In clinical practice, scintigraphy is usually

✉ Rina R. Rus
rina.rus@kclj.si

[1] University Children's Hospital, University Medical Centre Ljubljana, Bohoriceva 20, 1000 Ljubljana, Slovenia

[2] Department of Paediatric Nephrology, University Children's Hospital, University Medical Centre Ljubljana, Bohoriceva 20, 1000 Ljubljana, Slovenia

performed to rule out other CAKUT, especially renal ectopy and when renal compensatory hypertrophy is absent.

In the paper of Krill et al. [7], a direct comparison of US and scintigraphy (DMSA/MAG3) in diagnosing CSK was made on a cohort of 25 children. The results suggested that US alone is sufficient to diagnose CSK.

The aim of our retrospective study was to test on a larger cohort of children if US alone carries sufficient precision for a definite diagnosis of CSK.

Methods

A retrospective study was conducted at the University Children's Hospital in Ljubljana, Slovenia, and was approved by the Slovene National Medical Ethics Committee.

Patients

The medical charts of 310 children, diagnosed as having solitary kidney in the period from January 1980 to December 2017, were reviewed. Among them were 45 cases in which solitary kidney was due to the surgical removal of renal malignancy or dysplastic kidney, another 127 children who had functional solitary kidney with remaining contralateral kidney tissue confirmed by US, thus leaving us with 138 children having CSK with no contralateral kidney tissue found. In the last mentioned group there were 39 children who did not undergo renal scintigraphy and 99 children who met the inclusion criteria, namely, abdominopelvic US with no evidence of renal tissue on one side (empty renal fossa, no renal tissue within the retroperitoneum or pelvis) and confirmatory renal scintigraphy using either a DMSA or MAG3 scan, who were further analysed in this study.

We collected the following data: gender, side of CSK, number of prenatally abnormal US, age at time of first postnatal US, causes for the first US, US appearance of kidney and length measurements, additional CAKUT, urinary tract infection (UTI), extra renal anomalies and causes for scintigraphy.

Ultrasound

Abdominopelvic US was performed by different operators in supine positioned children using a 3.5–5 MHz probe mostly with one of Toshiba US machines (SSA 140A, Eccocee, Power Vision 6000, Ecusson). In all 99 cases, the first US showed one absent kidney, suggesting CSK. The compensatory hypertrophy of CSK was defined in prone positioned children as the kidney length on the maximum longitudinal kidney section greater than the value of the 95th percentile for a normal kidney length as described by Akhavan et al. [10]. US was considered normal when there was normal echogenicity, structure and thickness of kidney parenchyma, normal corticomedullary differentiation without calyceal dilatation, and 10 mm as an upper limit of the normal anterior–posterior renal pelvic diameter [11].

Scintigraphy

All children underwent renal scintigraphy with either DMSA or MAG3 to confirm CSK or to rule out obstruction or parenchymal scarring.

DMSA scintigraphy was performed 4h after intravenous application of radiotracer technetium-99m (99mTc). Images were recorded in supine position in posterior and posterior oblique views and an eventual additional anterior view; pinhole images were recorded in small children [12]. MAG3 scintigraphy was started immediately after injection of the bolus of intravenous radiotracer 99mTc in supine position. In the case of radiopharmaceutical lagging behind, the investigation was completed by intravenously injecting furosemide (0.5 mg/kg) during the investigation [13]. The minimal doses for 99mTc-MAG3 and DMSA were 17 MBq with adjustment of dose according to weight and actual dose card, as the dose card had been slightly modified in 2007 [14]. The children were not sedated during the DMSA and MAG3 scans, and parents were advised to hydrate their children well prior to the scans.

The results of the study were compared by calculating the accuracy of US vs. scintigraphy in the diagnosing of CSK. The value was calculated using the accuracy equation.

Results

Children with CSK were predominantly male (61.6%), and CSK was largely right-sided (55.5%).

The reasons for performing US, which revealed CSK, were as follows: abnormal prenatal renal US in 10/99 (10.1%) children, US after urinary tract infection in 8/99 (8%), US as part of examinations for other congenital malformations, enuresis, urinary incontinence or abdominal pain in 32/99 (32.3%) children; in 32/99 (32.3%) children US was performed in some hospitals as part of neonatal screening. In 17 children, the reason for the first renal US was not identifiable from the medical charts. The median age of children with a CSK diagnosis confirmed by US was 8 months (0.7 years: range 0–232 months; mean 4.0 ± 5.2 years).

In 41/99 (41.4%) children, US showed a normal kidney with appropriate compensatory hypertrophy, 23/99 (23.2%) had a normal kidney with kidney length between the 75th and 95th percentiles, and 8/99 (8.1%) had a normal kidney with kidney length under the 75th percentile. In 20/99 (20.2%) children, US was defined as normal or appropriate, but with no measurements recorded.

In the remaining 7/99 (7.1%) children, US was abnormal in terms of hydronephrosis, hydroureteronephrosis, structural changes in the parenchyma, and increased echogenicity.

CAKUT (other than CSK) was identified in 25/99 (25.2%) children, and more than one anomaly was found in 11/25 (44%) children. The most common anomaly was VUR in 15/99 (15.1%) children, followed by pielon duplex (4), hydronephrosis (3), hydroureteronephrosis (3), ectopic CSK (3), hydrourether (1), cystic dysplasia of CSK (1), uretero-pelvic junction obstruction (1), uretero-vesical junction obstruction (1), hypospadias (1), and detrusor sphincter dysinergia (1). In children with normal US appearance of CSK and kidney length above the 75th percentile (including those defined as appropriate with no measurement found), CAKUT was found in 17.8% (15/84) of children, compared to 66.7% (10/15) in the group with abnormal US and/or kidney length under the 75th percentile.

One or more UTI occurred in 27/99 (27.3%) children.

Extra renal anomalies (genital, heart, gastrointestinal, bone and inguinal hernia) were identified in 21/99 (21.2%) children, 13 of whom (13% of all) had a multiorgan syndrome (5 VATER/VACTERL association, 3 Rokitansky syndrome, 1 MURCS association, 1 Towns-Brock syndrome, and 3 undefined).

58/99 (58.6%) children underwent renal scintigraphy with DMSA and 41/99 (41.4%) with MAG3. In 97 cases, US correctly predicted the absence of (functional) renal tissue on one side in the renal fossa or in an ectopic location (pelvis or ipsilateral side). In two cases, scintigraphy was suspicious of renal tissue residues, which were not seen on US. In the first case, a MAG3 scan showed radiopharmaceutical activity on the empty side, which accounted for about 5% of the renal function (Fig. 1), and on subsequent US still no remains of the renal tissue could be found. In another case, a MAG3 test results showed the possibility of a very hypoplastic renal tissue that could be overlapped with the activity of the liver (Fig. 2). Although a DMSA scan was suggested, it was not performed due to normal renal function of the remaining kidney.

The calculated accuracy value of the abdominopelvic US in diagnosing CSK was therefore 98%.

Discussion

For a definitive diagnosis of CSK, renal scintigraphy is suggested as being the gold standard of ruling out ectopic functioning renal tissue, possibly missed by US [5, 12]. However, Krill et al. [7] have already proposed US as being sufficient to make a CSK diagnosis, stressing there was no need for a child being unnecessarily exposed to scintigraphy ionizing radiation. In light of their findings albeit on a small group of patients, we studied ultrasonography precision in comparison with renal scintigraphy on a larger cohort of children with CSK in order to determine if US alone is accurate enough to diagnose CSK.

Comparing our results to some other studies [1, 15], we found some differences in CKS size, the number of additional CAKUT, and extrarenal anomalies. All these differences could possibly be attributed to the fact that the diagnosis of CSK is established due to various reasons (abnormal prenatal US, UTI, abdominal pain, enuresis, urinary incontinence, searching for additional anomalies).

58/99 (59%) children had a scintigraphy scan using DMSA and 41/99 (41%) using MAG3 with similar time distribution over the years. DMSA scintigraphy is still regarded as the gold standard of ruling out ectopic functioning renal tissue and for checking the function of the solitary kidney [12]. However, according to Othman et al. [6], the MAG3 scan provides adequate information for assessment of the renal cortex and measurement of the split renal function, provides information on urodynamic, avoids unnecessary radiation, and is time saving.

In our study, US correctly predicted the absence of functional renal tissue on one side in the renal fossa or in an ectopic location (pelvis or ipsilateral side) in 97/99 cases (98%). The accuracy of US in diagnosing CSK is in agreement with the study made on a smaller cohort by Krill et al. [7], reporting 96% accuracy. We also agree with their observation that US technology development is progressing with streamlined resolution and parallel increase in the qualification of radiologists, so even more accurate achievements can be expected in the near future. The studies exhibit an evident increase of renal growth in the majority of CSK [7, 15], with overgrowth initiated prenatally [16–18]. Insufficient compensatory renal hypertrophy hints at a missed ectopic kidney [17]. According to literature, in less than 5% of all renal ectopias, an ectopic kidney could be found even intrathoracically [19], and as such, in the absence of any associated respiratory symptoms, easily overlooked. In our study, there was no (functional) intrathoracic ectopic kidney seen by scintigraphy, but non-functional kidney tissue could be overlooked, while systematic radiologic imaging of the thorax was not performed in all children due to a retrospective study.

Our results showed that in CSK patients having normal US with appropriate renal compensatory hypertrophy and no other problems (UTI, VUR, hydronephrosis, arterial hypertension or urinary incontinence in girls), renal scintigraphy does not add additional relevant information over US. It is necessary to determine the presence of a small ectopic kidney in case of other problems, such as arterial hypertension, UTI or urinary incontinence in girls, and an appropriate investigating procedure for a specific problem should be performed. Scintigraphy could be regarded as an essential next step in the diagnostic evaluation of girls with

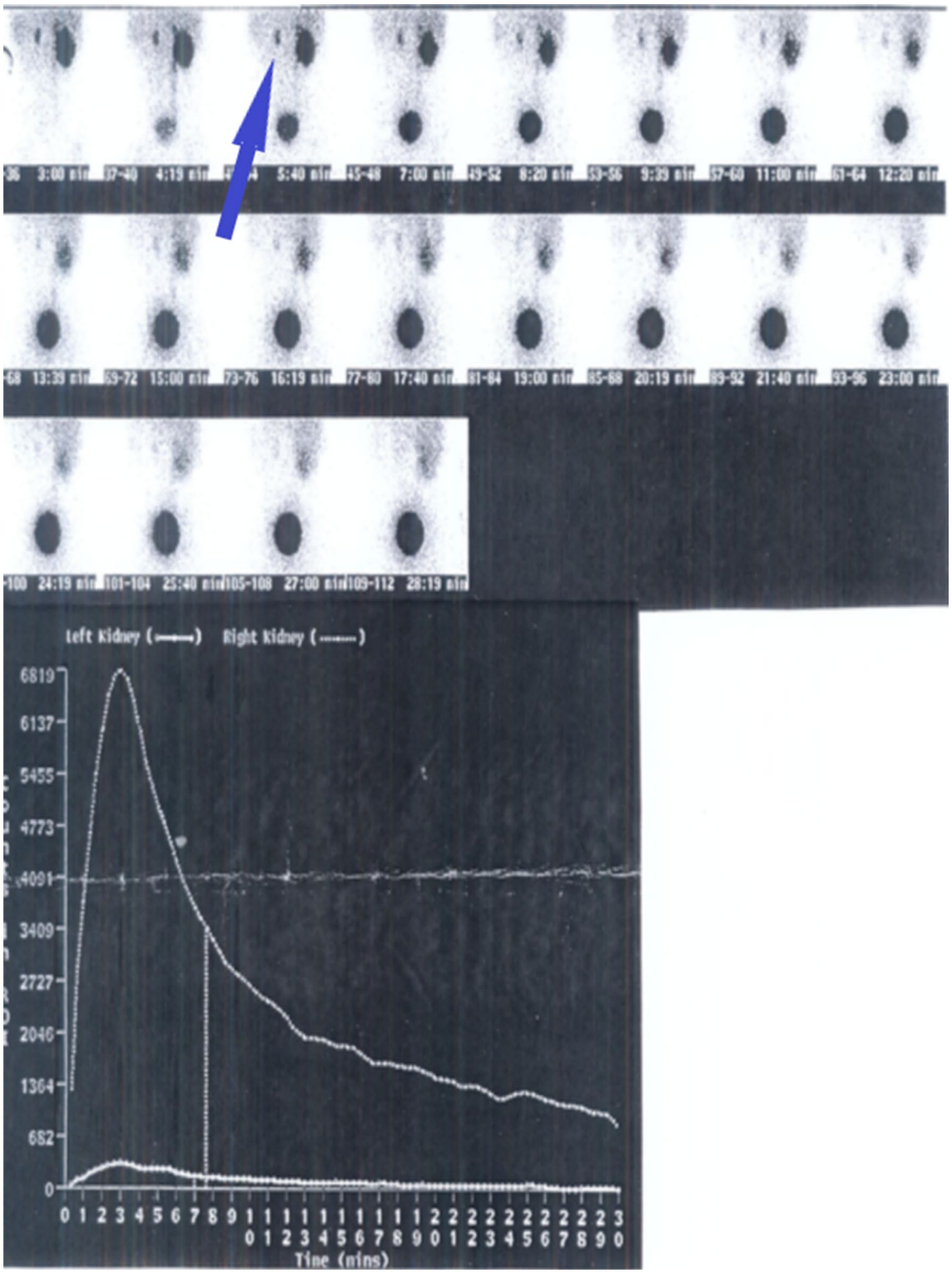

Fig. 1 Clearly visible kidney tissue (arrow) with 5% function on MAG3 scan on the side where on US no remains of the renal tissue could be found

continuous urinary incontinence and single kidney on US [20], although functional magnetic resonance urography (fMRU) should be the method of choice for depicting or ruling out an ectopic ureter [21].

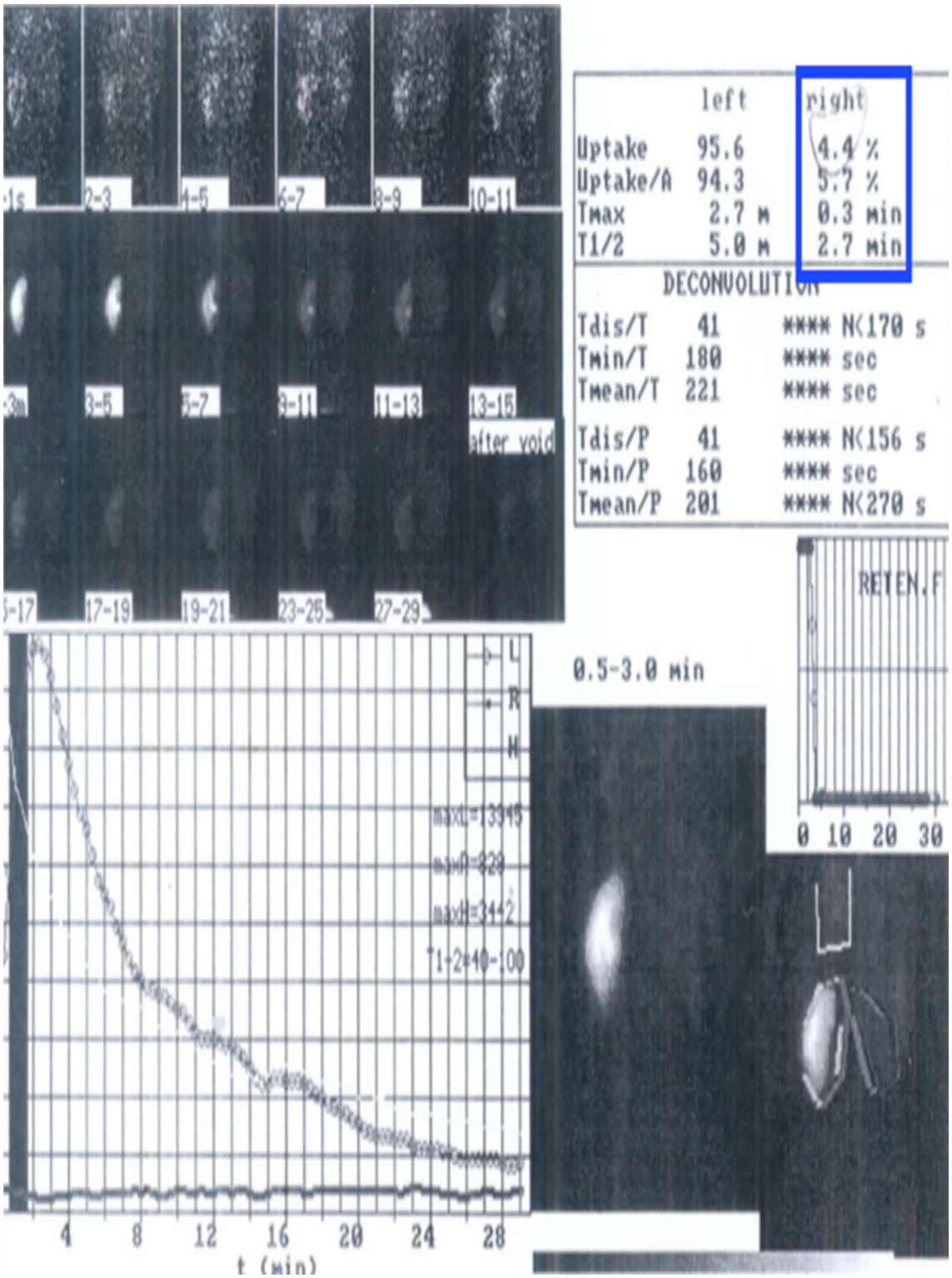

Fig. 2 MAG3 scan detected 4.4% activity on the side where no renal tissue was found on US, which allowed the possibility of a very hypoplastic renal tissue. Further diagnostics with DMSA scan was suggested

We suggest that routine confirmatory renal scintigraphy is not needed in CSK patients with normal US, with appropriate renal hypertrophy, and without other problems. Our proposed diagnostic approach, which includes US alone, appears to be accurate enough and more cost-effective, non-invasive, and child friendly than the standard one.

Conclusion

Our findings confirm that abdominopelvic US alone is accurate enough to diagnose CSK. If renal abnormalities are sonographically excluded and the patient has no other problems, routine confirmatory nuclear scanning may not be necessary. In this way we would ameliorate patient discomfort, avoid exposure to ionising radiation, and save money.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval A retrospective study was conducted at the University Children's Hospital in Ljubljana, Slovenia, and was approved by the Slovene National Medical Ethics Committee. A study is in accordance with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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