



Original Article

POSTNATAL MANAGEMENT OF PRENATAL OBSTRUCTIVE HYDRONEPHROSIS

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Introduction

Hydronephrosis is detected by prenatal ultrasonography. This method of surveillance detects a significant fetal anomaly in 1% of pregnancies, of which 20-30% of cases are genitourinary in origin, and 50% manifest as hydronephrosis^{1,2,3}.

Pelviureteric junction (PUJ) obstruction is by far the most common cause of paediatric hydronephrosis, occurring in 1 per 1000-2000 newborns.

Neonatal hydronephrosis has received significant attention since prenatal ultrasonography became a mainstream screening tool; however, the management postnatally remains controversial.

It has been suggested that patients with renal function better than 40% are monitored with repeat renal scans at 3- to 6-month and 12-month intervals, and surgery is performed only when a clear deterioration in renal function is present.

In this paper we advocate a different approach; waiting until function deteriorates is a less optimal option, especially when all the investigations to measure renal function are crude tests, for example diuretic renography, and even more important is the fact that loss of „renal reserve“ with the ongoing deterioration of kidney function is irreversible and unmeasurable.

We propose the use ultrasound in combination with renal scintigraphy to differentiate those infants who

would benefit from an early surgical intervention, highlighting the need to have a multi data point, jigsaw puzzle-like approach.

Eggshell sign

An additional test that allows us to differentiate those who made need surgery is the egg-shell sign. The ultrasound egg-shell sign (Figure 1) is seen as a thin crescent of increased echogenicity over a distended calyx. It is thought to be equivalent to the IVP Dunbar sign (Figure 2), which is a rim of contrast at the junction of the calyx and medulla in an obstructed distended kidney⁴.

The eggshell sign was first described in an abstract in 1999⁵, followed by the publication of a single case in detail^{6,7}, then the subsequent publication of a case series⁸.

Egg-Shell Sign Series:

To further evaluate the role of this additional sign a series of 94 patients with known obstructive pathology were reviewed. The sets of preoperative ultrasound images (not the reports) were compared to the clinical and radiological outcome, including comparing a review of the images with the outcome. In the 40 postnatal and 54 prenatally detected hydronephrosis cases, the egg-shell sign was seen in 25.0% of 16 diagnosed postnatally with urethral obstruction (COPUM)⁹, while in the 24 with pelviureteric junction obstruction the echogenic pericalyceal crescent was seen in 20.8%. Of the prenatal group, eight had urethral obstruction, 62.5% of whom demonstrated the egg-shell sign, whereas the prenatally diagnosed patients with pelviureteric junction obstruction displayed the egg-

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shell sign in 31 of 44 (70.5%). The results, and subsequent experience, seem to confirm that antenatally diagnosed hydronephrosis, due to significant urinary tract obstruction, is associated with the egg-shell sign on ultrasound, particularly in those patients with pelviureteric junction obstruction. Those patients without significant calyceal distension, and those with renal dysplasia or low pressure, severe hydronephrosis, do not display the egg-shell sign.

While PUJ obstruction diagnosed on antenatal ultrasound appears to have a strong association with the egg-shell sign, raised pressure may occur transiently in the fetal kidney because of the high fluid throughput in an otherwise near normal kidney. The characteristically high fetal urine output alone may cause transient calyceal dilatation¹⁰, and abnormally increased urine output without obstruction has been associated with unilateral hydronephrosis. It is conceivable that the relative polyuria could result in raised intrapelvic pressure akin to obstruction, leading to the egg-shell appearance on ultrasound.

It is speculative as to the actual anatomical basis of this radiological phenomenon. Possible explanations are the sonographic appearance of the interface between dilated calyx and adjacent parenchyma (hence the requirement for morphologically normal, non-dysplastic kidneys for production of the sign). Interestingly, a similar calyceal crescent sign has been described in excretory urography. The "Dunbar sign", which describes short, crescentic, opacified lines adjacent to dilated calyces, seen during intravenous urography in cases of high pressure hydronephrosis due to pelviureteric and vesicoureteric junction obstruction, may represent dilated and reoriented collecting tubules (4, 11) Given the similar morphological appearance, and possible association with obstructive hydronephrosis, the egg-shell sign is probably the sonographic equivalent of the Dunbar phenomenon.

Egg-shell Sign Index Case:

The initial index case was the male fetus of a 35 year old woman who had a routine ultrasound scan at 19 weeks gestation, which identified a left perirenal urinoma, minimal left renal parenchyma on aspiration of the urinoma, and right hydronephrosis. The right renal pelvicalyceal dilatation increased at 31 weeks gestation, associated with a rim of increased echogenicity adjacent to a number of the dilated

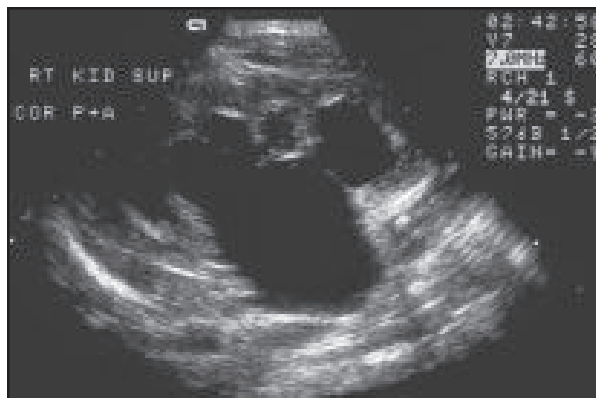


Fig.-1: Egg shell sign; a rim of echogenicity at the interface of the calyx and parenchyma is demonstrated in this coronal view of an obstructed kidney.

calyces. The child was delivered at 32 weeks gestation and was confirmed to have a right PUJ obstruction from the combination of an ultrasound within 12 hours that showed persistence of the calyceal dilatation, the egg-shell sign (Figure 2) and loss of renal corticomedullary differentiation, a rising creatinine, a nuclear medicine study showing poor renal function



Fig.-2: IVP Dunbar sign which is a rim of contrast at the junction of the calyx and medulla in an obstructed distended kidney

and limited drainage, and clinically increasing hydronephrosis and tension in the kidney. These factors resulted in an antegrade pyelogram and intrarenal pressure measurement that showed a raised intrarenal pressure greater than 30cm H₂O. Subsequently, findings at operation were of an inflamed and thickened renal pelvis and a PUJ obstruction, culminating in a pyeloplasty. Post operatively, the egg-shell sign and hydronephrosis resolved, although the crescentic rim persisted after the hydronephrosis was first seen to be settled at 10 days post-operatively.

Calyx to Parenchyma Ratio in Pelviureteric Junction Obstruction

The concept of the egg-shell sign has added to our ability to differentiate patients into management subgroups. In particular it seems to improve our ability to determine the degree of urgency for intervention for those with more severe changes. However, further data points, in other words, pieces of the jigsaw puzzle, seem necessary in some of those cases that are not straight forward. Thus, we have suggested the evaluation of the calyx to parenchyma ratio (Figure 3).

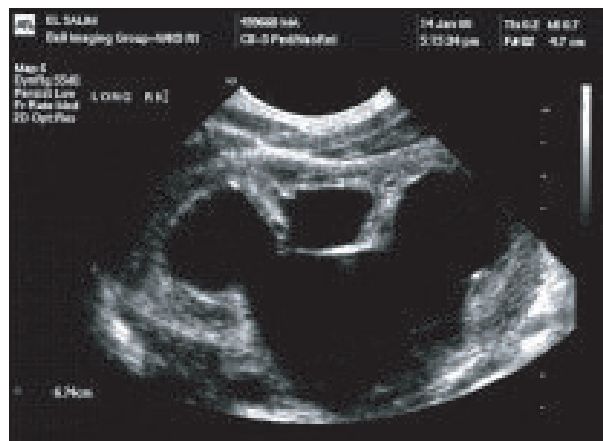


Fig-3: PUJ shows the deep calyces and the thin parenchyma reflecting a high CP ratio

A retrospective study of 58 PUJ obstruction patients was conducted, in which pre-operative and post-operative US were analyzed, the ratio of the depth of calyces to the thickness of the parenchyma (C/P ratio), and the operative and nuclear medicine findings correlated. The series showed a significant difference between pre-operative and post operative C/P ratios ($p < 0.01$), with good correlation between C/P ratios and the results of diuresis renography, showing the CP ratio to be a further, additional indicator of the

appropriateness of surgical intervention in renal obstruction.

Discussion

The ultimate goal of management of obstructive uropathy is to preserve renal function and avoid unnecessary surgery. Managing asymptomatic patients is challenging, because the natural course of hydronephrosis, due to obstruction, is variable.

Because there is no reliable method to predict whether the function of a kidney with obstruction will remain unaffected or deteriorate with time, there is controversy on the role of watchful observation with surgical intervention only if there is a deterioration in the patient's clinical status (i.e. decrease in renal function and/or development of symptoms).

Neste *et al.*¹² and Kis *et al.*¹³ suggested that hydronephrosis may be worse post operatively a contention that had not been reviewed systematically, and contrary to the commonsense expectation that an operation designed to improve drainage would decrease the degree of dilatation. This probably relates to the limited criteria used to evaluate post operative changes, using the Society of Fetal Urology Criteria, namely: 0 - normal kidney, 1 - slightly dilated renal pelvis without caliectasis, 2 - moderately dilated renal pelvis with mild caliectasis, 3 - large renal pelvis and dilated calyces with good renal parenchyma and 4 - very large renal pelvis with grossly dilated calyces and thin parenchyma.

Kis *et al.*¹³ showed the improvement in pelvic dilatation was quite slow in the first postoperative year while the increase in thickness of renal parenchyma is relatively fast. However, monitoring the size of the renal pelvis following Anderson-Hynes pyeloplasty has less meaning as the residual renal pelvis size is a reflection of surgical technique, particularly when trimming of the pelvis is performed.

Ransley *et al.*¹⁴ in their study of neonatal hydronephrosis did not focus on the details within the diuretic renogram study, nor did they approach evaluation of the ultrasound multifactorially. Patients with hydronephrosis with a clearly obstructed pattern on initial nuclear medicine scan are more likely to have poor final renal function and less likely to have normalization, with a significant risk of renal impairment, despite close monitoring, with the predictable adverse findings being confirmed at surgery by seeing all modalities as important to the

understanding of any one patient^{15,16,17}. We would suggest that the nuclear medicine study be reviewed concurrently with renal ultrasound to establish an algorithm to assist in the decision as to whether surgery is appropriate. Neither study can be viewed in isolation when deciding on surgery, and each study must be interpreted with multi data-points.

In the second phase of diuretic renography at peak renal uptake, intravenous Frusemide is administered and the excretion of isotope from the kidney is measured referred to as the “washout curve”. This phase indicates the extent of obstruction, if present. In the normal kidney, the administration of Frusemide results in a prompt washout. In a dilated system, if washout occurs rapidly after diuretic administration (<15 minutes), the system is not obstructed. If washout is delayed beyond 20 minutes, the pattern is consistent with obstructive uropathy. However, a delayed washout must be interpreted with caution^{18,19}. As an example, in a series of 39 infants with antenatal unilateral hydronephrosis followed without surgery, diuretic renography indicated obstruction in 24 patients whose renal function never decreased and thus could not have been obstructed¹⁹. These results may be in part due to the normally low neonatal glomerular filtration rate that can be refractory to diuretic therapy. If washout is between 15 and 20 minutes, the study is indeterminate. This is where we suggest using Ultrasound as a further screening tool, if the infant had the egg-shell sign prenatally and an increased C/P ratio; this indicates an element of obstruction that unlikely to resolve with observation. On the other side of the coin, it should be remember that adequate renal drainage on a diuretic renogram does not exclude intermittent obstruction. An observation of importance is the presence or absence of pain during the investigation and the comparison of the history of the presentation with the description of what happened for the patient during the study.

From the more surgical perspective, it is also worth noting that, when the C/P ratio is increased, or there is the egg-shell sign, the improvement in the appearance post surgery is greater if the surgery is conducted in the neonatal period, in line with the study by King *et al.* who strongly supported neonatal pyeloplasty, based on their series showing greater improvement in renal function in patients who undergo surgery before the age of 3 months²⁰. Chacko *et al.*²¹ demonstrated that early correction of PUJ obstruction

results in better drainage and a high chance of return to a normal configuration of the pelvicalyceal system. To wait for loss of function seems inappropriate, particularly as it has been shown that glomeruli are lost long before the glomerular filtration rate (GFR) is affected²².

Conclusion

Our previous study results, the literature and our subsequent experience, confirm that antenatally diagnosed hydronephrosis, due to significant urinary tract obstruction, is associated with the egg-shell sign and an increase in the C/P ratio on ultrasound, and that determining those cases that should be operated on early is as important as avoiding surgery on those who will come to no harm without intervention. We advocate the jigsaw puzzle approach to diagnosis to data collection on each patient, so as to avoid the Lasix clearance and pelvic dilatation two point decision making process.

We would suggest that the nuclear medicine study be reviewed concurrently with renal ultrasound to establish an algorithm to assist in the decision as to whether surgery is appropriate

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