Clinical-Radiological management of congenital hydronephrosis.

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Authors: M. Vidal, D. Llanos, E. Pallares, I. de la Pedraja, J. Arrazola; Madrid/ES
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Learning objectives

To review the main causes of congenital hydronephrosis (CHN) and its clinical and radiological management.
Background

Dilatation of the urinary tract is a common matter of concern in newborns.

It’s usually diagnosed before birth by ultrasound (US), but further imaging studies might be required during the neonatal period to assess its severity and choose the best options of treatment or follow-up in each case.

Severity ranges from mild and transient to severe.

Depending on the etiology, its severity and evolution, the patient will undergo a conservative follow-up or surgery. Sometimes prophylactic antibiotics are given to prevent infection during the follow-up.
Findings and procedure details

Babies who have shown a renal pelvis >4mm before the 33rd week of pregnancy or >7mm after the 33rd week should undergo a postnatal evaluation. The visualization of dilated calyces is always an abnormal finding, and these neonates need also to be imaged after birth.

Ultrasound is the initial technique to study the urinary tract of the neonate. It gives a superb anatomic detail, it's safe and widely available. Unless there is a suspicion of urethral obstruction, bilateral CHN or unilateral in patients with only one kidney, the first US imaging should be after the first week of life. Before this time, the degree of hydronephrosis can be underrated, because of the physiologic oliguria of the newborn.

The most accepted ultrasonographic grading system of the severity of CHN is the one proposed by the Society for Fetal Urology (Fig. 1 on page 6). This classification is based on the morphology of the pelvis and the calyces, which is way more important than the absolute value of the diameter of the renal pelvis.

There are five grades, from 0 to IV. The higher the grade, the more severe the hydronephrosis.

- Grade 0 is normal (Fig. 2 on page 6).
- Grade I and II (Fig. 3 on page 7 and Fig. 4 on page 8) if asymptomatic, are managed conservatively and may require follow up.
- Grades III and IV (Fig. 5 on page 9 and Fig. 6 on page 10). Patients with these higher grades of CHN should be also examined with void cystourerthrography (VCUG), to rule out VUR and evaluate the urethra. Some patients may benefit from contrast-enhanced US if a follow-up is needed.

When a significative obstruction is observed, an isotopic nephrogram is needed to assess the renal function.

The main causes of CHN are, in decreasing order of incidence:

1. **Transient benign hydronephrosis** (Fig. 7 on page 11), which resolves spontaneously during the follow up.
2. **"Physiological" hydronephrosis** (eg. extrarenal pelvis) (Fig. 8 on page 12), which remains stable and without symptoms or parenchimal involvement during the follow up.

3. **Pyeloureteral junction (PUJ) obstruction.** (Fig. 9 on page 13) This is the most common condition that requires surgical correction. The pelvicureteral transition is narrow and produces obstruction.

4. **Vesico-ureteral reflux (VUR)** (Fig. 11 on page 15 and Fig. 12 on page 16). Children with VUR are prone to urinary infections. It may regress spontaneously as the child grows up. It remains disputed whether to prescribe antibiotic profilaxis or not. Some severe cases need surgical correction.

5. **Obstructive megaureter** (Fig. 13 on page 17 and Fig. 14 on page 18). The distal segment of the urether is narrow, and produces a significant obstruction. Many cases are mild and have a good outcome during the follow up. If the hydronephrosis progresses, surgery may be needed.

6. **Ureterocele** (usually associated with duplex kidney). The duplex kidney has two moieties and two ureters (Fig. 15 on page 19 and Fig. 16 on page 20). The ureter of the upper pole moiety has a lower and more medial than normal end in the bladder. It usually ends in an ureterocele and is obstructive. The ureter of the lower pole ends higher and more lateral in the bladder, and is often refluxing. Some ureteroceles are so large that they produce an urethral obstruction.

7. **Posterior urethral valves (PUV)** (Fig. 17 on page 21, Fig. 18 on page 22 and Fig. 19 on page 23). The presence of redundant mucosal folds in the posterior urethral that produce bladder outlet obstruction is a severe condition, almost exclusive of boys. It usually associates uni or bilateral CHN, VUR or urinoma, and oligoamnios during the pregnancy. The oligoamnios may cause pulmonary hypoplasia and lead to death. It’s considered a surgical emergency.
Fig. 1: Figure 1. Classification of the Society for Fetal Urology for CHN.

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**Fig. 2:** Figure 2. Grade 0. Renal pelvis with normal morphology. No calyces.

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**Fig. 3:** Figure 3. Grade I: Rounded pelvis. No calyces.

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Fig. 4: Figure 4. Grade II. Rounded pelvis. Some calyces are visible, mildly dilated, but they hold a normal morphology. If it looks like a calyx, then it has a "normal" morphology!

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Fig. 5: Figure 5. Grade III. Dilated pelvis and calyces. The calyces are dilated and rounded (they no longer look as a calyx, but as a ball!). Grade III should undergo VCUG and isotopic nephrogram.

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Fig. 6: Figure 6. Grade IV. Severely dilated pelvis and calyces, with thinning of the renal parenchyma. The distorted morphology of the collector systems looks like a ghost for the authors. And even more scary!!!.

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**Fig. 7:** Figure 7. Grade II HN in a 7-day-old newborn. Another US was performed 6 weeks after, and the kidney looked normal then. This case could be referred as transient benign HN of the newborn.

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**Fig. 8:** Figure 8. Grade I HN in a 7-day-old newborn. Another US was performed 6 weeks after, and the pelvis remained slightly dilated (no calyces). Extrarenal pelvis ("physiologic" HN).

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**Fig. 9:** Figure. 9. Follow-up US in a child who showed grade II HN when one-week-old. Six weeks later, he has a grade III HN. Distal ureters were not dilated. The VCUG didn’t show VUR. PUJ stenosis.

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Fig. 10: Figure 10. Pyeloureteral junction (PUJ) obstruction

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Fig. 11: 1-day-old boy. Bilateral severe CHN on fetal US. The US scan showed grade III bilateral HN. Distal ureters were dilated and the VCUG showed high grade bilateral VUR. The urethra looks normal.

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**Fig. 12:** Figure 12. Vesico-ureteral reflux (VUR)

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Fig. 13: Figure 13. Grade III HN in a 1-week-old child. The distal ureter is dilated, with tapering of its lower end. The VCUG is normal. Obstructive megaureter.

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Fig. 14: Figure 14. Obstructive megaureter

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**Fig. 15:** Figure 15. Duplex kidney. There are two independent moieties in the right kidney. The upper one is dilated, and the renal parenchyma looks dysplastic. There's an ureterocele in the bladder and VUR to the lower moiety.

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**Fig. 16:** Figure 16. Duplex Kidney.

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Fig. 17: Figure 17. PUV Newborn with gross dilatation of both kidneys. Oligoamnios during pregnancy. Grade IV HN (fear the ghost!). Both ureters and the posterior urethra are dilated.

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**Fig. 18:** Figure 18. PUV. The VCUG shows bilateral VUR and dilatation of the posterior urethra.

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Fig. 19: Figure 19. Posterior urethral valves (PUV).

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Conclusion

CHN is a common finding. An accurate management is needed to achieve a good clinical outcome and avoid unnecessary imaging studies.

All newborns with an abnormal prenatal ultrasound must undergo a postnatal US evaluation. The best time to perform this US is at seven days of life. Children with bilateral CHN or suspicion of urethral obstruction must be US-scanned as soon as possible.

The morphology of the collecting systems and the thickness of the renal parenchyma are more important than the diameter of the renal pelvis. Pelvic diameter is useful to standardize screening criteria and for the follow-up of a same patient.

SFU grades> II require VCUG and isotopic functional study to know the etiology and assess renal injury. Asymptomatic (no UTI) children with grades #II are followed up with US and discharged when stability or resolution of the CHN is confirmed.

Only 10-20% of all the children with CHN seen in-utero will need surgery.
References