Antenatal Hydronephrosis and PUJO, Pelvi-Ureteric Junction Obstruction

Dr Clarence Lei Chang Moh,
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Advanced Urology Course (Paediatric Urology), HKL
OBSTRUCTIVE UROPATHY

Important as it may be reversible

Avoid renal failure if both sides

*Unilateral hydro not so urgent* !!
VUR

NEUROPATH
(SPINE, CNS, LL)
nonneuropathic

PUJO

VUJO

PUV

(BOY, bil)
Antenatal Diagnosis Dilemma
INTRA-UTERINE SURGERY
FETOMATERNAL TEAM –
detailed US, amniocentesis,

HYSTEROTOMY – few centres

Percutaneous – internal SPC, risk infection, miscarriage vs lung mature
DIAGNOSIS OF OBSTRUCTION

Palpable kidney, ureter, bladder
Ultrasound, within few days PP, 1 week, 1 month, 3 months

good exam. of anatomy
cannot assess true function
Mickey Mouse
PUJO us antenatal
Antenatal Hydronephrosis
What’s an Abnormal Ultrasound?

Society of Fetal Urology (SFU)

Hydronephrosis Grading Scale

Grade I
Grade II
Grade III
Grade IV

SFU scale applies to UPJ dilation not UVJ
Table 1  The relative importance of urinary tract malformations as a cause of lethal fetal anomalies (from 357 terminations of pregnancy with validation by autopsy and cytogenetics; (from [4])

<table>
<thead>
<tr>
<th>System</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central nervous system</td>
<td>171 (48%)</td>
</tr>
<tr>
<td>Urinary tract</td>
<td></td>
</tr>
<tr>
<td>bilateral renal agenesis</td>
<td>10</td>
</tr>
<tr>
<td>bilateral multicystic dysplastic kidney</td>
<td>9</td>
</tr>
<tr>
<td>unilateral MCDK/contralateral agenesis</td>
<td>3</td>
</tr>
<tr>
<td>infantile polycystic</td>
<td>1</td>
</tr>
<tr>
<td>outflow obstruction</td>
<td>10</td>
</tr>
<tr>
<td>dysplasia</td>
<td>1</td>
</tr>
<tr>
<td>complex</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>35 (9.9)</td>
</tr>
<tr>
<td>Complex</td>
<td>30 (8.5)</td>
</tr>
<tr>
<td>Chromosomal</td>
<td>25 (7)</td>
</tr>
<tr>
<td>Cardiac</td>
<td>23 (6.4)</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>14 (3.9)</td>
</tr>
<tr>
<td>Gastro-intestinal</td>
<td>7 (2)</td>
</tr>
<tr>
<td>Pulmonary</td>
<td>6 (1.7)</td>
</tr>
<tr>
<td>Tumour</td>
<td>1 (.03)</td>
</tr>
<tr>
<td>Twin related</td>
<td>2 (0.8)</td>
</tr>
<tr>
<td>Miscellaneous (unknown aetiology)</td>
<td>41 (11.5)</td>
</tr>
</tbody>
</table>

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Table 3  The final urological diagnosis in 426 live-born infants with significant prenatally detected uropathy (DFM Thomas, unpublished data)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pelvi-ureteric obstruction</td>
<td>150 (35.2)</td>
</tr>
<tr>
<td>Vesico-ureteric reflux</td>
<td>83 (19.5)</td>
</tr>
<tr>
<td>Multicystic dysplastic kidney</td>
<td>64 (15)</td>
</tr>
<tr>
<td>Vesico-ureteric junction obstruction</td>
<td>42 (9.8)</td>
</tr>
<tr>
<td>Posterior urethral valves</td>
<td>37 (8.6)</td>
</tr>
<tr>
<td>Duplex systems</td>
<td>36 (8.4)</td>
</tr>
<tr>
<td>Renal agenesis</td>
<td>14 (3.3)</td>
</tr>
</tbody>
</table>

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Prenatally diagnosed hydronephrosis: the Great Ormond Street experience

H. K. Dhillon
The Department of Paediatric Urology, The Hospital for Sick Children, Great Ormond Street. London, UK

Introduction

Prenatal ultrasonography (US) has identified a large population of fetuses with a urinary tract abnormality that is most commonly a non-specific hydronephrosis. Such a dilatation will eventually be found to be associated with an upper or lower urinary tract 'obstruction', reflux or duplex kidneys. However, almost half of all prenatally diagnosed hydronephroses are found after investigation to be an isolated upper tract dilatation consistent with the radiological diagnosis of a PUJ obstruction. This population has posed the greatest challenge in prospectively determining which of these asymptomatic children have an obstruction that would benefit from surgery, as opposed to a simple dilatation that may be inconsequential to their well-being.

History

The advent of maternal US in 1980 led to an inappropriate enthusiasm for early surgery which was justified on the basis that a hydronephrosis with poor drainage during diuretic renography constituted a PUJ obstruction [1]. An unsubstantiated emphasis was therefore placed on the failure of tracer washout after administering furosemide during renography [2,3]. However, drainage curves obtained after administering furosemide are affected by many variables, e.g. the state of hydration of the infant, the function of the kidney and bladder emptying. The Great Ormond Street experience in the field of prenatally diagnosed hydronephroses evolved from understanding the necessity to elucidate the natural history of this population. In 1980, Ransley et al. proposed that drainage curves after furosemide were not reliable indicators of obstruction in this population and introduced instead the concept of using differential function, thereby extending the role of the renogram in this group [4]. A protocol based on using differential function for determining management was formulated and formed the basis of the first 'natural history' series in this new population [5]. The second phase of our experience involved the construction of a randomized clinical trial to compare the outcome of early surgery with conservative management in a group of children with prenatally diagnosed unilateral hydronephroses. Finally, the result of 17 years of managing a large population consistently according to protocol has contributed significantly to identifying the population at risk of renal functional deterioration or symptoms, as well as allowing those with an inconsequential dilatation to be followed with minimal imaging [6].

The 'natural history' series

This series comprised 115 children with an upper tract dilatation who were referred to our unit between 1980 and 1988. The group constituted consecutive referrals, excluding any child who was referred after 6 months of age. There were 148 hydronephrotic kidneys, of which 77 had unilateral dilatation with a normal contralateral kidney; the dilatation was bilateral in 33 children and five children had solitary hydronephrotic kidneys. The follow-up for the children in this series ranged from 5 to 16 years. The protocol introduced in 1980 was based on classifying all the hydronephrotic kidneys into one of three functional groups depending on the uptake of 99mTc-DTPA. A poorly functioning kidney was designated as having <20% of overall function, a moderately functioning kidney as 20–39% and good function as ≥40%. Those in the last group were clearly the most important, as the dilemma in this population was largely whether healthy children with good function in hydronephrotic kidneys actually had a PUJ obstruction which would benefit from surgery. All 148 kidneys in the series did not drain before the administration of furosemide but poor drainage did not influence either the classification or management decisions. The degree of hydronephrosis was recorded by measuring the anteroposterior diameter of the renal pelvis in the transverse plane of the kidney on renal US. The initial imaging comprised US in the first week of life to confirm the prenatal diagnosis. Functional imaging was postponed until 4–6 weeks of age and the differential function recorded then enabled the kidneys to be classified into one of three functional groups. On initial imaging, kidneys with poor function (<20%) proceeded to surgical intervention. The group with moderate function (20–39%) and good function (≥40%) were re-imaged at the age of 3 months with
Is a dilated system always obstructed?

No

It may be due to:

a) congenital, nonobstructive
b) reflux
c) previous obstruction
d) “physiological” postpartum, diuresis esp antenatal & neonatal, full bladder
IVU, 6 WKS, lasix
Megaureters

Defined as a dilated ureter

Types

1. Refluxing
2. Nonrefluxing nonobstructed (Primary megaureter)
3. Obstructed
- Study of anatomy, not function
- delayed drainage at tight stricture is suggestive
IVU

(a) Delayed excretion
(b) Prone view
(c) P. M.
(d) Post-lasix

X-rays poor measurement of function
Poor drainage may be due to capacity effect esp with poor fn
OBSTRUCTED, NOT DILATED

1. Decompressed, eg urinoma
2. Dehydrated
3. U/S obscured
4. Intermittent
RADIO-ISOTOPE Tc DTPA

- Quantifies (a) function (GFR) (b) excretion ($t^{1/2}$)

- BUT, poor excretion may be from Capacity effect of dilated system, esp if poor fn
(Lower third of ureters ROI)

14722 MAX

(2) ureter (distal third)

Right ureter (distal third)

6

150.0 SEC./DIV
SERIAL RADIO-ISOTOPE

- deterioration of absolute function may indicate obstruction

- BUT, need to rule out other causes of deterioration

*(WASHOUT less important cf Fn)*
Definition of Obstruction

- Obstruction exists in a fluid-transporting system if the fluid pressure proximal to a relative narrowing must be raised to transmit the usual rate of flow through this area.

(RH Whitaker, personal communication)
WHITAKER’S TEST

Intrapelvic pressure
(eg needle or nephrostomy)

Perfusion at 10 ml/min
> 22 cm H2O : obstructed
15 – 22 : equivocal
< 15 : unobstructed

Why 10 ml/min?
Leakage around nephrostomy
TRIAL OF DECOMPRESSION

- usu nephrostomy
- ureteric stent

Significant improvement of function (in the absence of other factors) confirms obstruction.
Open, lap, robot: how stents, remove, NT, CBD

Other MIS – success?  

Followup – US, IVU/isotope, 5 yrs
SGH
Robot
Assisted
6 cases, 2008,
BW 10.8 kg, 2yrs
Cases
Thank you