CONGENITAL HYDRONEPHROSIS: PROBLEMS IN DIAGNOSIS AND MANAGEMENT

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ABSTRACT

Objective: *To review problems in diagnosis and management of congenital hydronephrosis.* ***Material & methods:*** *This study was retrospective. Data were collected from medical records of patients with congenital hydronephrosis, which were hospitalized or consulting the urologic outpatient clinic at Ciptomangunkusumo Hospital from January 1999 to December 2008 and Harapan Kita Maternal and Pediatric Hospital from January 2004 to December 2008. Data were analyzed with SPSS programme version 13.0. Statistical analysis was performed to find the relationship between age at diagnosis and kidney function (Mann-Whitney test) and between age at diagnosis and nephrectomy rate (Chi-Square test).* ***Results:*** *Antenatal ultrasound was performed in only 59 out of 63 patients and only 44,07% (26 patients) with hydronephrosis was detected antenatally. Eleven out of 26 antenatally diagnosed patients came to our clinic at a later age. Three standard studies (postnatal ultrasound, voiding cystourethrogram, and renal scintigraphy) were performed only in 12 out of 145 patients (8,27%). Ultrasound was performed in 108 patients (74,5%), voiding cystourethrogram in 79 patients (54,5%), and renal scintigraphy in only 26 ptients (17,9%). The suggested management was conducted in 115 patients; operative management in 95 patients (82,61%) and conservative treatment in 20 patients (17,39%). The most common operative procedures were ureteroneocystostomy, pyeloplasty, nephrectomy, and posterior urethral valveablation. Mean serum creatinine in the below 12 months old group and above 12 months old group was 0,78±0,93mg/dl dan 1,03±0,88 mg/dl respectively($p<0,05$). There was no significant difference in nephrectomy rate in both age groups ($p>0,05$). Nephrectomy was performed in 16 patients, in which the most common indication was grade IV hydronephrosis with thin parenchyma in 11 patients (68,75%) and the most common etiology was UPJ obstruction in 10 patients (62,5%). We could only collect follow up data from 73 out of 115 managed patients (63,48%). Urinalysis, ultrasound/voiding cystourethrogram, and renal function studies were not routinely conducted during follow up.* ***Conclusion:*** *The management of congenital hydronephrosis in Indonesia needs improvement in antenatal care standards, particularly obstetric ultrasound, to improve early detection of congenital hydronephrosis. Better education for parents about the importance of follow up is needed, especially for antenatally diagnosed patients. A consensus regarding diagnostic tools used in managing congenital hydronephrosis should be established among urologists, pediatricians, and radiologists.*

Keywords*: Congenital hydronephrosis, diagnosis, management.*

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INTRODUCTION

Hydronephrosis is defined as abnormal dilatation of pyelum and/or calices, which is often associated with renal parenchyma changes.[1] If hydronephrosis is already present or diagnosed before the baby is born, it is called antenatal or congenital hydronephrosis. A multicenter study in Europe (EUROCAT working group) found that the prevalence of congenital hydronephrosis is 11.5 out of 10,000 live births.[2]

Congenital hydronephrosis is the most common anomaly found in antenatal ultrasound examinations,[3,4] and accounting for 50% of all

congenital anomalies.[5] Early detection of hydronephrosis is very helpful in postnatal management. Causes of congenital hydronephrosis are ureteropelvic junction (UPJ) obstruction, multicystic kidney disease, vesicoureteral reflux, double collecting system, ureterovesical (UVJ) junction obstruction, and posterior urethral valves.[6]

Patients diagnosed with congenital hydronephrosis in the antenatal period should be re-evaluated with postnatal ultrasound to confirm hydronephrosis; voiding cystourethrogram (VCUG) to see presence of vesicoureteral reflux, vesical anomalies, and to exclude possibility of posterior urethra valves; and renal scintigraphy to evaluate significant obstruction.[6-8] Follow up with serial ultrasound is still needed because obstruction in some patients can be found later in life. VCUG examination is recommended in all congenital hydronephrosis patients, including patients with known resolution of hydronephrosis because 14% of these patients still have vesicoureteral reflux,[9] and 45% of them will have urinary tract anomalies.[10]

In Indonesia, diagnosis and management of congenital hydronephrosis is still a problem. Antenatal ultrasound examination has not played its maximum role in detecting congenital hydronephrosis. Unnecessary diagnostic examinatons are often done. Another problem is the limited availability of renal scintigraphy examination in medical centers in Indonesia. Diagnostic problems are associated with further management problems, i.e. which patients need surgical intervention and which patients only need conservative management.

OBJECTIVE

The aim of this study is to evaluate diagnostic and management problems in congenital hydronephrosis.

MATERIAL & METHODS

This study is a retrospective study of all congenital hydronephrosis patients in Urology Clinic or hospitalized in Department of Urology Ciptomangunkusumo General Hospital between January 1999 December 2009 and Harapan Kita Women and Children Hospital between January 2004 December 2008. Data is processed with SPSS 13.0. The results were analyzed using Mann-Whitney test to find out relationship between age at diagnosis and renal function; using Chi-Square test to find out relation between age at diagnosis and nephrectomy rate.

RESULTS

There were 145 patients with congenital hydronephrosis with median age 29 months and ranges between 1-183 month, and consisting of 55 females and 90 males. In this study, 62 patients had renal function deterioration based on increased serum creatinine (refers to normal value of serum creatinine based on children age group).

In this study, 15 of 145 patients came because they were already diagnosed with congenital hydronephrosis during antenatal period; the rest came with symptoms such as abdominal mass, urinary retention, recurrent urinary tract infection, urinary incontinence, or symptoms of decreased renal function. Sixty three out of 145 patients had their antenatal care history explored through phone interview. Table 1 shows antenatal care and antenatal ultrasound characteristics in those 63 patients.

Table 1. Characteristic of antenatal care and antenatal ultrasound of patients with explored antenatal care history.

No.	Antenatal care	Amount of patients (%)
1.	Caregiver	
	Obstetrician	56 (88,89%)
	General practitioner	1 (1,59%)
	Midwife	6 (9,52%)
2.	Antenatal Ultrasound	
	YES	59
	Detected hydronephrosis	26 (44,07%)
	Undetected hydronephrosis	33 (55,93%)
.	NO	4

Table 2. Etiology of congenital hydronephrosis.

Type of anomaly	Number of patients	Sex
UPJ obstruction:	40	32 males, 8 females
Unilateral	31 (24 left, 7 right)	
Bilateral	9	
Vesicoureteral reflux:	37 (9 primary reflux, 24 secondary reflux, 4 unclear)	20 males, 17 females
Unilateral	15 (12 left, 3 right)	
Bilateral	22	
Neurogenic bladder	25	
UVJ obstruction:	20	14 males, 11 females
Unilateral	14 (9 left, 5 right)	12 males, 8 females
Bilateral	6	
Double collecting system:	20	4 males, 16 females
Unilateral	18 (11 left, 7 right)	
Bilateral	2	
Posterior urethral valve	18	
Bladder neck contracture	13	6 males, 7 females
Ectopic ureter	10 (8 left, 2 right)	9 males, 1 females
Ureterocele	8 (5 left, 3 right)	3 males, 5 females
Anterior urethra anomaly	4	2 females with urethral stenosis, 2 males with meatal stenosis
Proximal ureteral stenosis	1	Female
Unestablished diagnosis	19	12 males, 7 females

Of 26 patients who had been diagnosed congenital hydronephrosis since antenatal period, 11 patients went to Urology Clinic at an older age, i.e. 2 patients at age 3 months, 3 patients at age 4 months, 1 patients at age 4.5 months, 1 patients at age 5 months, 1 patients at age 10 months, 2 patients at age 2 years, and 1 patients at age 8 years. Delay in visiting doctor is caused by parental factors who assumed that their children have no symptoms or were afraid that their children will undergo surgery.

There are several examinations for etiology of hydronephrosis as shown in Table 3. In this study, only 12 patients underwent all 3 recommended examinations (ultrasound,VCUG, and renal scintigraphy). In this study, ultrasound examination did not routinely assess the anteroposterior diameter of pyelum.

Table 3. Diagnostic modality.

Modality	Amount of patients
Ultrasound	**108 (74,5%)**
VCUG	**79 (54,5%)**
KUB-IVU	66 (45,5%)
Urethrocystoscopy	63 (43,4%)
CT Scan31	(21,4%)
Retrograde pyelography	29 (20,0%)
Renal scintigraphy	**26 (17,9%)**
Antegrade pyelography	7 (4,8%)
MRI	4 (2,8%)
Urethrography	3 (2,1%)

Out of 145 patients, 115 patients underwent recommended management, i.e. 95 patients underwent surgery (82,61%) and 20 patients (17,39%) underwent conservative management. The rest (30 patients) have not done follow up visit hence the management is not clear yet.

Table 4. Type of surgery done for congenital hydronephrosis.

Type of surgery	Amount of patients	Amount of intervention	Note
Ureteroneocystostomy was	16	20	In 4 patients, this intervention done both side.
Pyeloplasty	17	17	
Nephrectomy	16	16	
Posterior urethral valve ablation	15	15	
Bladder neck incision	12	12	In 5 patients, the incision was done at the same time with other intervention.
Nephrostomy (indefinite yet)	10	10	In 4 patients, an intervention was Also done in contralateral kidney
Endoscopic incision of ureterocele	7	7	
Vesicostomy or open cystostomy	4	4	
Heminephrectomy	2	2	
Excision and anastomosis of ureter	1	1	
Ureteral orifice incision, transureteroureterostomy, continent urostomy	1	1	
External urethral orifice incision	1	1	
External urethral orifice dilatation with clamp	1	1	

Note: more than one procedure could be done in one patient, for example: posterior urethral valve ablation and bladder neck incision or bladder neck incision and ureteroneocystostomy.

Table 5. Mean Serum creatinine and nephrectomy rate.

Variable	< 12 months age group	> 12 months age group	p (Statistic test)
Mean serum creatinine (mg/dL)	0,78±0,93	1,03±0,88	p<0,05 (Mann-Whitney)
Nephrectomy rate (n/%)	3/46 (6,52)	13/99 (13,13)	p>0,05 (Chi-Square)

Table 6. Etiology of congenital hydronephrosis and indication of nephrectomy.

Varible	n (%)
Etiology of hydronephrosis in patients who underwent nephrectomy	
UPJ obstruction	10 (62,5)
UVJ obstruction	2 (12,5)
Ectopic ureter and double collecting system	4 (25,0)
Indication of nephrectomy	
Stage IV hydronephrosis, renal enlargement, thin cortex	11 (68,75)
Non functioning kidney from renal scan	3 (18,75)
Renal hypoplasia	1 (6,25)
Contracted kidney	1 (6,25)

Serum creatinine rate and nephrectomy rate in less and more than 12 months age group can be seen in Table 5. Mann-Whitney test showed significant difference between those two age groups (p<0,05). Chi-Square test showed no significant difference in nephrectomy rate between those two age groups (p>0,05).

Table 7. Type of examination done during follow up.

Type of examination	Surgically managed (%)	Conservatively managed (%)	Total (%)
Urine analysis			
- No urinalysis/urine culture	46 (70,77)	5 (62,50)	51 (69,86)
- UTI (+)	16 (24,6)	1 (12,50)	17 (23,29)
- UTI (-)	3 (4,61)	2 (25,00)	5 (6,85)
Ultrasound/VCUG during follow up			
- No data	38 (58,46)	2 (25,00)	43 (58,90)
- Hydronephrosis/reflux improves	13 (20,00)	1 (12,50)	14 (19,18)
- Hydronephrosis/reflux persists	12 (18,46)	2 (25,00)	14 (19,18)
- Hydronephrosis/reflux deteriorates	2 (3,08)	0 (0,00)	2 (2,74)
Renal function assessment during follow up			
- No data	55 (84,61)	7 (87,50)	62 (84,93)
- Function improves	5 (7,69)	0 (0,00)	5 (6,85)
- Function deteriorates	5 (7,69)	1 (12,50)	6 (8,22)

*) Amount of patients who renal function assessed is very low and most came for follow up visit in early post-intervention stage.

Out of 115 patients who underwent management, only 73 patients came for follow up visit (63,48%). Type of examination done during follow up can be seen in Table 7. Out of 95 patients who underwent surgery, 65 patients (68,42%) came for follow up visit, and 27 of them (41,54%) underwent ultrasound/VCUG. The stage of hydronephrosis improved in 13 patients; persisted in 12 patients; and deteriorated in 2 patients.

DISCUSSION

Detection of congenital hydronephrosis by antenatal ultrasound was first reported in 1979 since Kay et al reported two cases of hydronephrosis detected during fetal period.[11] Since then, antenatal ultrasound played a great role in screening patients who need postnatal reevaluation and management.

In this study there are 145 patients with congenital hydronephrosis; 55 females and 90 males. This finding is in accordance with previous reports mentioned that the ratio between males and females is 2-4:1.[2,7,12] This study finds that only 15 patients out of 145 came because of antenatal diagnosis. The remaining patients came because of symptoms.

Two factors causing low percentage of patients who came in early stage postnatal are antenatal caregiver and parents. In 59 patients who underwent ultrasound examination in antenatal period, there are only 26 who had been diagnosed as hydronephrosis. This shows that obstetric ultrasound examination has not been optimally used in detecting congenital anomalies, especially urinary tract anomalies. Parental reluctance against bring their children to visit physicians after birth also contributes in delayed management of congenital hydronephrosis.

The consequences of delayed diagnosis are seen in the significant amount of patients who present with decreased renal function (42,76%) and elevated serum creatinine level in >12 months age group ($p<0,05$). Percentage of patients who underwent nephrectomy in >12 months age group is also higher than in < 12 months age group, although the difference is not statistically significant ($p>0,05$). Chertin et al (2002) stated that patients with congenital hydronephrosis and definite signs of obstruction who underwent release as early as possible, there is higher possibility of maintaining renal function and hence nephrectomy can be avoided.[13]

The gold standard diagnostic modality to find etiology now is postnatal ultrasound, followed by voiding cystourethrogram (VCUG), and renal scan (renal scintigraphy). There are several opinions on the best time to re-evaluate with postnatal ultrasound. The

most recent agreement is to perform ultrasound examination at >48 hours age because the first 48 hours is a transient oliguric phase in neonates hence hydronephrosis especially in mild stage could not be seen.[8] Postnatal ultrasound is done soon after birth in neonates suspected in antenatal period with posterior urethral valve, bilateral dilatation, solitary kidney, and oligohydramnion.[14] Which patients need conservative therapy and which need surgery can be predicted through those three modalities. Dhillon (1998) associates anteroposterior diameter or pyelum with incidence of surgery in patients with UPJ obstruction. He found that in patients with anteroposterior diameter 15-30 mm, 30-50 mm, and >50 mm, the incidence of surgery is 38%, 75%, and 100%, respectively.[15] VCUG examination is recommended for all patients with congenital hydronephrosis, including patients in whom hydronephrosis resolved at birth because vesicoureteral reflux is still detected in 14% of these patients.[9] Renal scan should be performed in all patients with congenital hydronephrosis to evaluate presence of significant obstruction. Renal function < 40% and decrease of renal function >10% in two renal scans are the indications for surgery.[16,17]

In cases of congenital hydronephrosis managed conservatively, serial ultrasound examinations play a big role in patients' follow up. Although initial renal scan (renal scintigraphy) does not show significant obstruction, but in some patients, obstruction can happen later in life. Serial ultrasound examination during follow up visits in patients on conservative management is a must.[18,19]

Problems that appeared in this study was inconsistency in application of standard diagnostic modalities to evaluate etiology of hydronephrosis. Gold standard examination were not done on all patients (ultrasound only to 74,5%, VCUG to 54,5%, and renal scan to 17,9% patients). Only 12 patients (8,27%) underwent all three recommended examinations. Ultrasound examinations did not mention anteroposterior diameter of pyelum. There is a significant amount of patients who underwent non standard examinations such as KUB-IVU (45,5%) and CT scan (21,4%).

From this description, we can see that ultrasound plays an important role in postnatal management of congenital hydronephrosis patients. To determine which patients should undergo conservative or surgical management, especially in medical centers without renal scan facility, ultrasound can be a pivotal tool.

From a management perspective, 82,61% patients had surgery and 17,39% had conservative management. Onen (2007) described that most congenital hydronephrosis cases can be managed non-operatively except with evidence of obstructive injury. In Onen study, 228 renal units with UPJ obstruction from 162 neonates were observed. He found that 201 renal units (88,2%) had spontaneous resolution with non-operative approach and only 27 renal units (11,8%) need pyeloplasty. Close follow up with periodic ultrasound screening for hydronephrosis and periodic renal function assessment with renal scan can help to identify cases which need surgical intervention to prevent permanent loss of kidney function.[17]

Out of 115 patients managed, only 73 (63,48%) came to follow up visits. This showed low parental awareness of importance of following up patients with hydronephrosis regardless of management. Examinations such as urinalysis, re-ultrasound/VCUG, and renal function assessment are not routinely done in patients who came to follow up. Ultrasound is a non-invasive diagnostic tool which is very sensitive, accurate, and easy to use, can be used to assess improvement or deterioration of hydronephrosis stage.[12,18,20]

Out of 27 post-operative patients who had been examined with ultrasound/VCUG, 13 (48,15%) had improvement in hydronephrosis stage; 12 patients (44,44%) unchanged; and 2 patients (7,41%) deteriorated. This fact shows that surgical intervention in congenital hydronephrosis is beneficial in improving hydronephrosis stage or preventing deterioration in hydronephrosis stage. Chertin et al (2002) reported that in patients with congenital hydronephrosis patients who underwent surgery because renal function deterioration, 36 out of 44

patients showed renal function improvement within 6-12 months postoperatively.[13] In this study, postoperative renal function assessment gave unsatisfactory result. This might be caused by only a few patients came to follow up visit and underwent renal function assessment, and those patients came in early postoperative stage hence renal function improvement could not be assessed yet.

CONCLUSION

The standard antenatal care needs improvement, especially obstetric ultrasound to detect congenital hydronephrosis; parents, especially informed of children's diagnosis in antenatal period, need to be educated about importance of follow up visits; and diagnostic modalities used (ultrasound, VCUG, and renal scan) need to be standardized between specialists who contributes in management of congenital hydronephrosis (urologist, pediatrician, radiologist). If those aspects are improved, then congenital hydronephrosis can be better managed, especially in determining which patients need surgical intervention hence reducing loss of renal function.

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