

# Acute Oligohydramnios: Antenatal Expression of VURD Syndrome?

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## Key Words

Fetal bladder outlet obstruction · Oligohydramnios · Renal function impairment

## Abstract

**Objective:** Oligohydramnios (OA) is nowadays regarded as one of the best markers of renal function (RF) impairment in bladder outlet obstruction (BOO) detected in utero. As such, its onset is usually early and progressive because of decline in fetal urine production. A series of acute OA complicating pregnancies with BOO has never been reported. **Methods:** Over a 7-year period, 5 fetuses with in utero suspicion of BOO exhibited an abrupt decrease of amniotic fluid after the 30th week of gestation. **Results:** All fetuses were delivered by cesarean section: diagnosis was posterior urethral valves in 3 cases, urethral atresia in 1, and prune-belly syndrome in 1. Urologic work-up demonstrated a unilateral vesicoureteral reflux dysplasia (VURD syndrome) in all 5 fetuses. RF at 1 year was normal in 4 fetuses and impaired in 1. **Conclusions:** Besides obstetrical reasons, OA may also have acute onset occurring in the presence of anomalies of the urinary tract; although diagnosis is almost always BOO, functional and anatomical characteristics of the urinary tract are those of VURD syndrome with a non-functioning, refluxing renal unit. The

associated acute OA/VURD syndrome may represent a milder expression of a pop-off mechanism advocated in this syndrome with a more favorable prognosis than progressive OA detected early in pregnancy.

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## Introduction

The relationship between amniotic fluid (AF) and renal and urinary tract anomalies (UTA) has long been established: at about 22 weeks, AF is formed primarily from fetal urine (7–10 ml/kg/h) and eliminated mostly by fetal swallowing up to 1 l/day [1]. Not surprisingly, a large number of papers have taken into consideration AF as a pronosticator of renal function (RF) [2]. While it is generally agreed upon that oligohydramnios (OA) is one of the best determinants of poor RF after birth [3], such considerations mostly apply to cases of bladder outlet obstruction (BOO) where renal involvement is bilateral. However, there is as yet no consensus as to the best measurement or threshold as a predictor in this pathology or as to whether gestation of onset has a role [4]. Nevertheless, OA in UTA is usually early and progressive, i.e. by the second trimester of pregnancy when the 'amniotic fluid

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**Table 1.** Perinatal characteristics of the 5 patients and RF at 1 year

Case	Maternal gravity/parity	AFI <5th percentile weeks GA	Karyotype (prenatal)	Birth weight g	Diagnosis (postnatal)	Grade V VUR		Scr (1 year)
						unilateral	bilateral	
1	2/1	31	46,XY	2,950	PUV	yes	no	0.5
2	1/1	31	46,XY	2,830	PUV	yes	no	0.7
3	1/1	34	46,XY	2,960	PUV	yes	no	0.4
4	1/1	34	46,XY	3,180	UA	yes	no	1.5 <sup>1</sup>
5	1/1	37	46,XY	3,000	PBS	yes	no	0.6

Scr = Serum creatinine (mg/dl); PUV = posterior urethral valves; UA = urethral atresia; PBS = prune-belly syndrome.

<sup>1</sup> Patient with Scr 1.5 = UT diversion.

cycle' is installed [5], the main source of production is the fetal kidney, with the fluid being subsequently cleared through the placenta after deglutition and absorption from the gastrointestinal tract [6]. On the other hand, acute late-onset OA has been repeatedly described in the literature for obstetrical reasons [7]. However, there are no published series examining relationships between such acute-onset OA and urinary tract abnormalities such as BOO.

### Patients and Methods

A database of all patients seen over a 7-year period for suspected BOO was reviewed. Inclusion criteria included: antenatal diagnosis (severe bilateral hydronephrosis; persistently distended bladder with or without visualization of dilated posterior urethra), diagnosis made between 17 and 20 weeks gestational age (GA) and acute, late-onset OA. For evaluation of AF, measurement by the amniotic fluid index (AFI) was considered [8]. All AF measurements were made by obstetricians with great experience in antenatal ultrasound. Maternal issues contributing to the OA such as medications or AF leak were investigated and excluded. All pregnancies with suspected BOO and intact AF were closely monitored at 7- to 10-day intervals: sudden increase was defined as normal AFI decreasing to <5th percentile at the next encounter. Late-onset OA was defined as AFI <5th percentile occurring after 30 weeks GA. All fetuses underwent formal karyotyping. Fetuses with intrauterine growth retardation, abnormal karyotyping, other structural anomalies and early OA were excluded as were those with stable AF throughout pregnancy. Fetal growth restriction was diagnosed on the basis of a birth weight <10th percentile for GA [9].

Clinical records of patients with late, acute-onset OA were then extracted and attention was directed to the following parameters: urologic diagnosis, RF (expressed as creatinine values) at 1 year of age and differential RF. Differential RF was assessed by nuclear scan.

### Results

Twenty-eight fetuses were seen during the period considered: of these, 5 met the inclusion criteria with acute-onset OA. In all cases, AF remained decreased throughout the rest of pregnancy and no changes in hydronephrosis were observed. The characteristics of the 5 patients with karyotype and RF at 1 year are shown in table 1. All fetuses were delivered by cesarean section at 37 weeks GA and survived. Once delivered, 4 patients voided spontaneously, while in 1 patient suprapubic drainage was positioned. Urine output was adequate in all cases. No patient required mechanical ventilation. Urologic diagnosis was made by cystourethrogram (performed transurethrally in 4 cases and from a suprapubic tube in 1 case) when the babies were stable. Posterior urethral valves (PUV) in 3 cases, urethral atresia in 1 case and prune-belly syndrome in the remaining 1 were diagnosed. In all patients a non-functioning, severely refluxing renal unit was found at further evaluation with nuclear scan (VURD syndrome).

### Discussion

Fetal kidney becomes the main source of AF by the second trimester of pregnancy, therefore in the vast majority of UTA, secondary to severe disturbance of anatomical and functional integrity of the kidney, onset of OA will occur at that time. Callen [10] has calculated that a fetus producing only 1 ml of urine per hour would result in halving AF in 3 weeks. In this respect, all papers taking into consideration variations of AF or in utero procedures with regard to future RF are mostly

focused on fetuses with early and progressive decrease of AF [11, 12].

When reviewing our series of BOO, we noted that in 5 cases AF declined abruptly and late (after 30 weeks GA). We were able to extract this small subset of patients since it is our policy to closely evaluate AF in fetuses with suspected BOO in collaboration between the obstetrician and urologist, therefore in our small series we were sure that OA was really of acute onset and not due to missing observations.

Acute OA is frequently associated with obstetrical problems such as premature rupture of membranes [13] or with drug administration during pregnancy [14]; the only mention in the literature of acute OA secondary to UTA is that of Noe [15] who reported acute OA in a fetus with prenatal ureteropelvic junction obstruction evolving into anuria and renal failure after birth. Differently from this observation, our patients did not require emergency postnatal diversion since RF was normal or improving – the only exception was a fetus with urethral atresia who is now in renal failure.

The only common feature of all these patients was severe monolateral reflux in non-functioning kidneys, in other words the so-called VURD syndrome. This syndrome was described in 1982 by Hoover and Duckett [16] as persistent unilateral reflux to a dysplastic non-functioning kidney in a boy with PUV. A pop-off mechanism, dissipating high pressures generated by obstruction into the dilated ureter, was called upon to explain such an association [17].

After this observation, numerous other reports appeared in the literature describing urinomas [18] and urinothorax [19] as expression of such a pop-off mechanism. However, the prenatal history of these conditions is limited to evaluation of AF only at the time of the observation prompting diagnosis. In this respect, all pregnancies with suspected BOO were followed by us with serial observations so that we could exactly determine the onset of OA. As previously mentioned, an association resembling VURD syndrome was found in all patients in this small series. Strictly speaking, VURD syndrome was described only for PUV, however the obstructive component is obvious in the cases of UA and can be considered functional in the prune-belly syndrome, so we felt such a definition could be extended to all cases.

Can there be any association between acute-onset OA and VURD syndrome? We speculated that sudden opening of the vesicoureteric junction, generated by excess pressure within the urinary tract exerted by BOO, may lead to AF backflow with 'gushing' of fluid into the extra

space provided by tortuous, hypoperistaltic dilated ureter, which actually becomes a reservoir for urine. Consequent disruption of the AF cycle could, in turn, be responsible for the acute OA which was observed in these fetuses. In other words, the sudden decrease in AF could represent an initial, mild expression of the postulated pop-off mechanism. Urinoma and urinothorax could reflect a later stage when pressure within the dilated urinary tracts exceeds the compensation potential.

Late-onset OA along with partial pressure relief within the urinary tract induced by the pop-off mechanism could explain normal RF observed at 1 year in 4 of our patients. Our series is too small to draw conclusions, neither can the two groups of patients with acute and chronic OA be compared statistically. An analysis of this issue in other types of congenital anomalies of the urinary tract is also lacking, however, we made an observational study on a small yet significant number of fetuses with acute OA where a VURD syndrome was demonstrated in all cases postnatally.

This finding once again outlines the importance of AF as a predictor of RF, however, late- and acute-onset OA may be a more favorable prognosis than an early occurring, progressive decline throughout pregnancy: the necessity of immediate admission and treatment of the newborn may not be necessary and parents can be somewhat reassured regarding quality of life. Therefore, when BOO is diagnosed prenatally, we recommend evaluation of AF to always be performed by experienced sonologists made aware not only of its importance in the follow-up of all UTA, but also of the necessity of conducting it until term.

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