One case of «Prune Belly» syndrome: prenatal and prognostic valuation

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Summary: A case of "Prune Belly" syndrome, its sonographic diagnosis, from the 15th week and its monitoring by sonography and biochemical exams of fetal urine for study of renal function is described.

The good relation between prenatal prognosis and neonatal renal function is verified after birth.

High resolution sonography available today makes feasible the study of the urinary system from the beginning of gestation.

Fetal kidneys are evaluable from the 10th week of gestation, even if better definition is possible later, when the middle structures of the organ are visible: cortex, medulla, renal pelvis and arcuate arteries ([1]).

The bladder is visible at the 13th week ([2]) and its morphology and dynamics can be studied by repeated checks every 30-60 minutes ([3]). Amniotic fluid volume and its variations are an index of the urinary system's functions ([4, 5]).

The initial routine sonographic checks (at the 16th and 18th weeks) can also visualize some primary morphologic anomalies or anomalies resulting from urinary system obstruction; other anomalies can be discovered later.

"Prune Belly" syndrome is characterized by the triad urinary system dilation, abdominal wall distension and cryptorchidism.

The term "Prune Belly" defined by Osler in 1901 ([6]), refers to the aspect of the abdominal wall which resembles a prune.

Skeletal, gastroenteric and kariotype abnormalities (13 and 18 trisomy and Turner syndrome) are frequently associated with the classic triad ([8]).

Our report concerns a case of a fetus affected by a "Prune Belly" syndrome, whose urinary system anomalies were discovered early (during the 15th week of gestation). We will discuss diagnostic and therapeutic procedures.

CASE REPORT

A 35 year old primipara, affected by ulcerative colitis diagnosed the year before, took salazopirina up to the sixth month of pregnancy.

During the first sonographic examination at the 15th week an abdominal anechogenic image 6 cm in diameter was visualized and another anechogenic elongated image was seen at its side,

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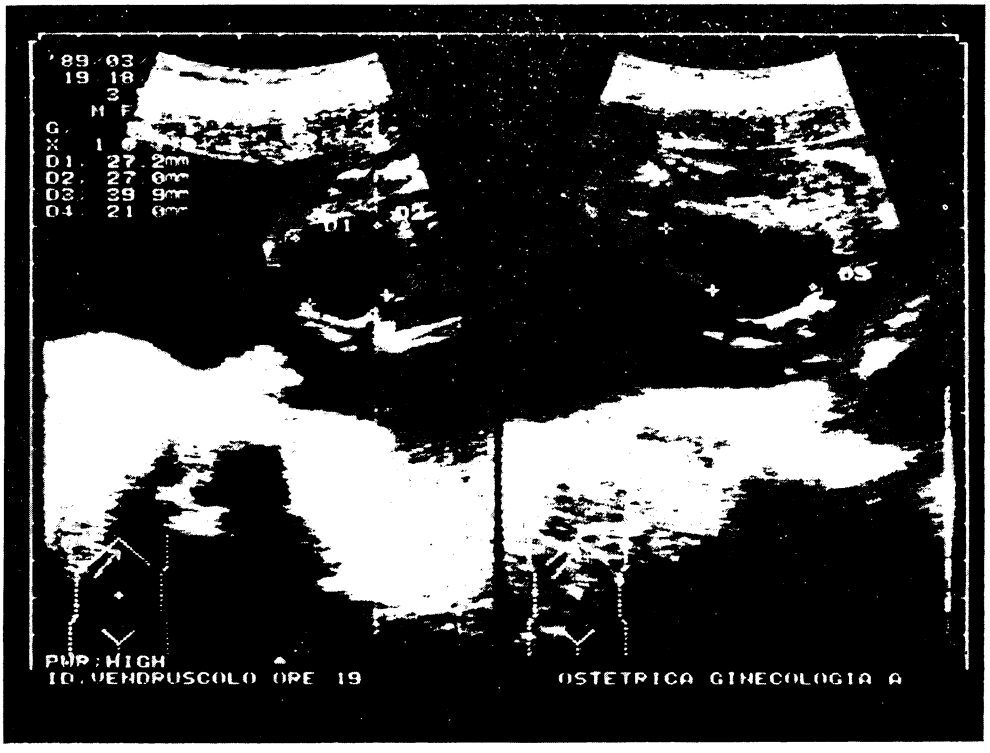

Fig. 1.

apparently not in communication with the first one.

The other morphological fetal aspects (growth parameters and amniotic fluid volume) were normal.

Blood examinations showed the presence of antibodies against Cytomegalovirus and Herpes Simplex 1 and a moderate anaemia.

At the 16th week of gestation an amniocentesis was performed to evaluate the fetal karyotype: the result was a normal karyotype 46 XY. A sonographically guided aspiration of the abdominal mass obtained 60 cc of clear fluid.

A few days later a sonographic check showed the same abdominal image, measuring 4 cm in diameter with a moderate ascites.

A following check at the 20th week showed a pear shape aspect of the mass (cm 3.5 in diameter), the absence of ascites and a good fetal growth.

At the 21st week the communication between the elongated mass and the pear shape mass was visible (photo 1) and another elongated anechogenic image appeared on the other side.

In the left kidney was visible a dilated pelvis and an initial hydronephrosis, while the right one was normal. The diagnostic hypothesis of bladder and ureteral dilation with initial monolateral hydronephrosis was confirmed.

Following checks of bladder emptying showed a negative result.

At 23rd week bladder and kidney dilation were unvaried. A fetal urine sample was taken to assess renal function, according to Glick and Harrison (9). The results obtained showed a good renal function (osmolarity 156 mosm, Na 160 mEq/ml, Cl 118 mEq/ml, proteinuria 22 mg/ml).

The fetal urinary samples was studied at the 25th week for the appearance of a right pelvis dilation. The typical abdominal wall fluctuation in the amniotic fluid that suggests the "Prune Belly" syndrome was also noticed in this period.

At the 28th week the bladder appeared more enlarged ($8.4 \times 6.7 \times 4$ cm); a further distention appeared after the 32nd week.

While the renal growth rate proceeded according to the 95° percentile, a progressive renal

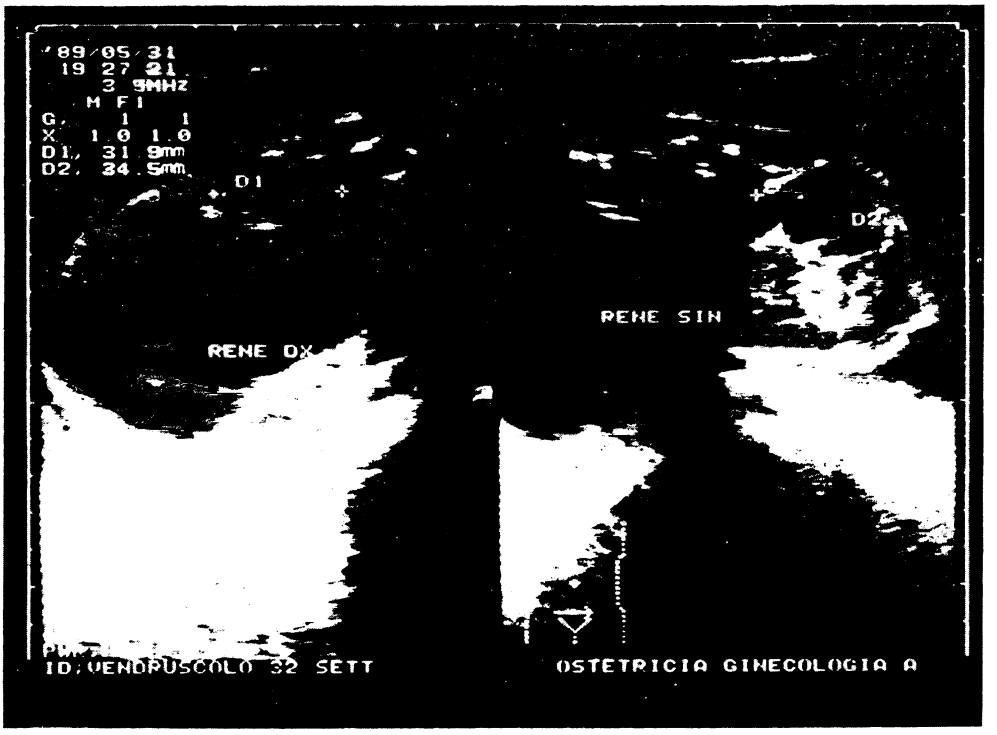

Fig. 2.

pelvis dilation was noticed in the left kidney, with a cortical thinning (photo 2).

No substantial changes appeared in the following weekly checks; the fetal growth curve remained normal.

At the 37th week a cesarean section was performed; the newborn was a male of 2900 g. (photo 3).

The newborn appeared with an evident abdominal musculature hypoplasia, cryptorchidism and clubfeet.

Neonatal sonography, urography, cystography and photoscintigraphy confirmed the presence of hydronephrosis in the right kidney with a thin cortex and a reduced renal function.

Urination was spontaneous with conspicuous bladder residue.

DISCUSSION

In our report this morphological anomaly was observed during the first routine sonography at the 15th week.

Even if the abdominal anomaly was singled out so early, the diagnosis of its nature was only made at 25th week, when the communication between the dilated ureter and the corresponding renal pelvis dilation were visualized.

Only at the 23rd week the typical abdominal wall fluctuation suggested the diagnosis of "Prune Belly" syndrome.

Also the literature review on this syndrome confirmed that the diagnosis is made around 20-23 weeks, suggested by the presence of a cystic abdominal mass, hydronephrosis and the abdominal wall anomaly ([9, 10, 11, 12]).

The frequent association of chromosomic anomalies and fetal structure abnormality imposes, as in this case, a study of the fetal karyotyping because the presen-

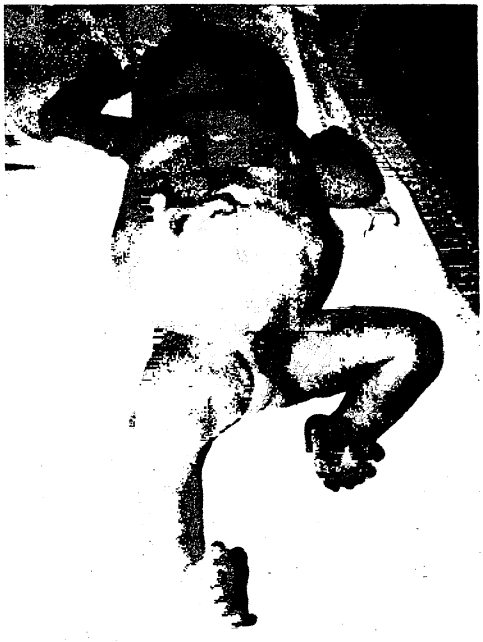

Fig. 3.

ce of these chromosomic anomalies changes the fetal prognosis.

In our case, with a normal fetal karyotype, we evaluated fetal "well being" considering these parameters: amniotic fluid volume, renal parenchimal ecogenicity, fetal urinary osmolarity and Na and Cl concentration.

We did not study the evaluation of the renal flow described by Glick [9], because we considered this procedure too complicated.

The three parameters we considered were sufficient for a good check of renal function.

We discussed the utility of the application of a vescico-amniotic shunt in correlation with the right renal pelvis dilation appearing at the 23rd week to avoid the increase of the obstacle to the urinary flow.

The normality of the urinary parameters, evaluated again at the 25th week, the volume of amniotic fluid and the normal echogenicity of the renal parenchyma persuaded us of the uselessness of the procedure.

The persistence of the favourable prognostic elements allowed us to wait for the physiological end of gestation.

The validity of these considerations has been confirmed by a study of the renal functionality performed after birth.

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