A CASE OF PRUNE-BELLY SYNDROME: PRENATAL DIAGNOSTIC PROBLEMS

C. GIGLI (*), A. AZZARITA (**),
G. D'OTTAVIO (*), M. A. RUSTICO (*),
G. P. MANDRUZZATO (*)

(*) Children Institute,
Obstetric and Gynecological Clinic,
University of Trieste (Italy)

(**) Obstetric and Gynecological Department

Ospedale Generale Provinciale, Schio (Italy)

SUMMARY

The Authors discuss a case of "Prune-belly syndrome" which could be diagnosed prenatally thanks to echography; they stress the importance of echographic controls since the first gestational weeks to better recognize pathological findings, and describe their attempts to drain the megavesica. However these attempts could not reverse the ominous prognosis.

The "Prune-belly Syndrome" (P.B.S.) or Prune-belly anomaly is a rare malformation affecting the urinary apparatus and the abdominal wall of the fetus.

Since 1895, 250 cases ([1]) have been reported in literature. The most recent of them were examined echographically too ([2, 3, 4, 5]).

P.B.S. affects mainly males (95%) ([6]) and is characterized by the absence of abdominal muscle, anomalies of the urinary apparatus and cryptorchidism.

Sometimes (postural?) changes in the lower limbs can also appear.

The anomalies, of varying seriousness, are believed to originate from an urethral obstruction and the consequent dilatation of the vesica, that can attain a really impressing size.

The compression exerted by the "megavesica" that tends to fill the whole of the abdomen, is believed to cause structural changes in the abdominal wall and lung hypoplasia, which accompanies this syndrome.

Thus, the neonatal death of these children appears to be due to respiratory and renal insufficiencies.

Consequently it is clear that the "megavesica" is the most characteristic element of this syndrome which is named after the typical aspect of the newborns when the "megavesica" is emptied, recalling a dried prune.

P.B.S. etiology is still unknown. The most recent hypotheses suggest the existence of hereditary transmission mechanisms, which could explain the higher incidence on males ([6]).

In this study we report on a case observed, for which prenatal diagnosis was possible thanks to echography, and on attempts of intrauterine decompression of the "megavesica".

CLINICAL CASE

L. C., age 35, primigravida, teacher.

She was sent to our institute after many echographic examinations performed in different

Clin. Exp. Obst. Gyn. - ISSN: 0390-6663
VIII, n. 2, 1981

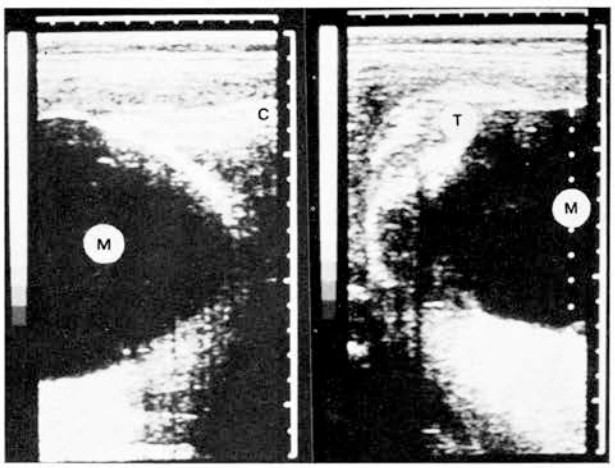
Centres, with the following diagnosis: "Suspected fetal ascites in primigravida at the 23rd week".

The patient's uterus was more developed than normally in relation to the period of amenorrhea (symphysis-bottom distance: cm 29). On its surface there were various myoma nodes.

The placenta stood on the bottom of the uterus, against the right wall.

Different hypotheses were examined. Some suggested the presence of a sac compressing the fetus from the outside; others the existence of a fetal sac, probably a "megavesica".

In order to clarify the situation, with the authorization of the patient, we performed a centesis of the intrauterine sac, under echoscopic control, and injected 20 cc of contrast medium (Bilivistan).



1 2

Fig. 1. — Fetal longitudinal echographic scanning. The "megavesica" (M) and the backbone (C) are visible.

Fig. 2. — Fetal transversal echographic scanning, just under the heart. To be noted: the fetal trunk (T) deformation and size, compared to the "megavesica" (M).

The echographic examination (Aloka SSD-250, probe 3.5 MHz) showed a really particular picture.

Most of the uterus was occupied by a large echotransparent sac, cyst-like, ovoid, about 12× 9.6 cm wide. It was impossible to measure its length exactly because of the probe size and the lack of reference points (figs. 1 and 2).

The fetus was compressed in podalic position wit hits back against the left uterine wall. The fetus head was normal; its biparietal diameter (cm 5.8) was regular compared to the amenorrhea period. Its backbone too was normal, whereas the thorax was reduced and deformed into a "sickle" shape, with the concavity, in profile, on the sac side.

Heart beating was normal. Abdominal organs were not clearly visible. The upper limbs were normal and showed slight movements, but the lower limbs were particularly far away from the pelvis and hardly visible, due to the retropubic position.

A small sac of liquid was located between the fetus head and the large described sac.

The radiologic picture did not prove very useful to the solution of the problem (fig. 3), nor did the tests performed on the liquid taken by centesis (tab. 1).

The fetal karyotype, subsequently performed on the cells of that same liquid, proved that of a normal male.

As a differential diagnosis between amniotic fluid and fetal urine, based on the biochemical dosages carried out on the liquid extracted, was impossible, we tried to obtain it from the liquid cells.

According to the anatomopathologist, these cells came from the amniotic fluid, whereas, according to the nephrologist, they came from the upper tract of the escretory ducts.

Repeated echographic observations, more than examinations, lead us to the convinction that it was most likely a P.B.S. case and that the most suitable therapy was the decompression of the megavesica, also considering the patient's firm determination to carry on her pregnancy.

of the cyst-like shape and the absence of inner echoes.

We were still in doubt between a "megavesica" and an ovarian cyst in a female fetus, as we had not yet the results of the fetal karyotype.

Considering that the size of ovarian cysts reported in literature is smaller than that of the examined sac, that they usually appear at later stages of pregnancy, and that, in this case, a serious oligoamnios was also present, we concluded that such a large sac of liquid could only originate from the vesica.

The particular deformation of the tho rax and the anomalous position of the lower limbs (already stressed by other Authors in connection with P.B.S.) supported our idea.

We therefore concluded that only P. B. S. could explain all the observed anomalies.

It is fair to admit that the other examinations, far from making a decisive contribution to the diagnosis, were hardly useful if not, in some occasions, misleading.

All attempts to drain the "megavesica" to reduce compression on the thorax and allow lungs to expand proved useless. This was probably due partly to the small quantity of urine extracted, compared to the whole mass, and partly to the probably irreversible stage reached by the fetus pathologic changes.

CONCLUSION

The echographic record of a considerable cyst-like sac of liquid, starting from the fetus abdomen, suggests the existence of a P.B.S.

The increasing size of this sac, in a series of observations, and the presence of an oligoamnios support this thesis.

A more timely diagnosis will be possible if echographic controls begin at an early stage of pregnancy.

BIBLIOGRAPHY

1) Ives E. J.: *Birth Defects Original Article Series, 10*, 127, 1974.
2) Garrett W. J., Kossoff G., Osborn R. A.: *Brit. J. Obst. Gyn., 82*, 115, 1975.
3) Okulski T. A.: *J. Clin. Ultrasound, 5*, 268, 1977.
4) Cooperberg P. L., Romalis G., Wright V.: *J. Association of Radiologists, 30*, 120, 1979.
5) Bovicelli L., Rizzo N., Orsini L. F., Michelacci L.: *Clin. Genetics, 18*, 79, 1980.
6) Riccardi V. M., Grum C. M.: *J. Med Genetics, 14*, 266, 1977.