Prune Belly Syndrome with Hydrocephalus and Craniostenosis

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Abstract

Introduction: The Prune Belly syndrome (PBS) is a complex and rare malformation with a clear male predominance, characterized by the association of a major bladder dilation, a distention with impaired muscles of the anterior abdominal wall that is flaccid and wrinkled and cryptorchidism. PBS is not a genetic disease.

Objectives: To make known this pathology through our observation.

Observation: This is a newborn male, born at home, eutrophic term resulting from a pregnancy not followed. Admitted to the neonatology unit at 2 hours of life. He had an abdominal protrusion whose musculature is loose; anal polyp; the left foot bot talus; cryptorchidism; the anterior fontanel closed. The exploration found craniostenosis, hydrocephalus with cerebral parenchymal hyper-echogenicity, nephromegaly with bilateral hydronephrosis, and mega-bladder. A multidisciplinary management (neuro-surgical, nephrological, urological, orthopedic and pediatric) was programmed for this child but the parents did not show up for the appointments, having required a convocation after which they inform us that their child is dead!

Conclusion: The diagnosis of Belly Prune Syndrome is easy from the 15th week of pregnancy; urologic surgery improves the prognosis and delays the occurrence of renal failure main cause of mortality. In 2019 there are many pregnancy not followed at our country.

Keywords: Prune Belly Syndrome; Newborn

Introduction

We had this case in 2009, but it did not have the opportunity to be published. It happened when I was doing my civil service in a mother-child establishment in the city of Oran, in Algeria.

The prune Belly syndrome (PBS) is a complex and rare malformation with a clear male predominance, characterized by the association of a major bladder dilation, a distention with impaired muscles of the anterior abdominal wall that is flaccid and wrinkled and cryptorchidism. PBS is not a genetic disease.

Objectives of the Study

To make known this pathology through our observation.

Our observation

This is a newborn male, born at home eutrophic term from a non-follow-up, admitted to the neonatology unit at 2 hours of life.

Citation: Hayat Aichaoui. “Prune Belly Syndrome with Hydrocephalus and Craniostenosis”. EC Paediatrics 9.7 (2020): 92-95.
Clinical examination at admission

- Weight 3 kg300, PC 32 cm
- Strong cries
- 136 bpm, no heart murmur
- 42 c/min, no respiratory distress
- Anal polyp, good passage of the rectal probe
- Protrusion of the abdomen, absence of abdominal musculature
- Small penis, purses are empty
- Left talus clubfoot
- Closed anterior fontanel
- The archaic reflexes are present and normal.

Para-clinical examinations

- HT: 45%
- Blood urea 0.12 g/l, serum creatinine 6.3 mg/l
- Negative CRP
- Radio chest probe in place and ASP are normal
- Trans-fontanellar echography: mega communicating big cistern with the ventricular system, moderately dilated V3 and V4, associated with a cerebral parenchymal hypergenesis.
- Abdominal echo: Enlarged kidneys with significant dilatation of pelvic-calyctic cavities bilaterally almost completely laminating the renal parenchyma Thin - walled mega - bladder discharges laterally and upwards the digestive loops. Spleen and liver of normal appearance, and absence of intraperitoneal fluid effusion.

At 24 hours of life

- Generalized mottling, lack of meconium and oligo-anuria
- Hypertonic convulsions of the limbs and perioral cyanosis Supported
- Sedation of convulsions by Gardenal® then related by depakine® after 48 hours
- Antibiotics
- Breastfeeding initiated after hemodynamic and neurological stabilization

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- Monitoring of urinary catheter diuresis, transit monitoring: rectal probe stimulation and evacuation lavement.
- An appointment was fixed for the follow-up at Uro-nephro-pediatric service.

**Evolution**

- Parents do not accept the illness of their child, despite our interview with them and the psychological care within our service.
- New born put out as soon as the feeding became satisfactory, and resumption of a normal diuresis, with a control in the 3 following days at the level of our consultation.

For a follow-up in parallel with the specialized care in the service of pediatric uro-nephrology and psychological care.

The parents did not show up for the appointment, but 2 months later they come back to inform us of the death of their child

**The usual treatment**

**Is surgical:**

- Urine diversion by nephrostomy, cutaneous ureterostomy, cystostomy, in severely affected patients
- Reconstitution of the urinary system by reimplantation and modeling of the ureter according to an anti-reflux procedure
- Treatment of cryptorchidism in several times
- Plication of the abdominal wall.

**Usual prognosis of the disease:**

- Most often lethal the first weeks of life
- Chronic constipation, severe abdominal meteorism
- Pulmonary hypoplasia
- Chronic urinary tract infections, renal lithiasis, kidney failure
- Infertility
- And other complications related to the associated diseases.

**Prevention**

Antenatal diagnosis is possible to 15 SA, with an antenatal surgical correction.

**Conclusion**

The diagnosis of Belly Prune Syndrome is easy from the 15th week of pregnancy, urologic surgery improves the prognosis, and delays the occurrence of renal failure main cause of mortality [1-4].

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Bibliography


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