New Sonographic Sign of Congenital Chloride Diarrhea: Can We Avoid Unnecessary Operations - 💊

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Submitted: 31 March 2020; Approved: 25 April 2020; Published: 27 April 2020

Citation this article: Zefov V, Abdelrady F. New Sonographic Sign of Congenital Chloride Diarrhea: Can We Avoid Unnecessary Operations. Open J Pediatr Neonatal Care. 2020;5(1): 004-006.

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INTRODUCTION

Chloride diarrhea is a rare form of congenital diarrhea caused by an autosomal recessive gene. A few hundred cases have been reported, mainly in Saudi Arabia, Finland and Poland. It is a serious bowel disorder due to electrolyte disturbances affecting the chloride/bicarbonate (Cl/HCO3) channels; intraluminal chloride concentrations are very high, with hypochloremia and hypokalemia, causing the patient to develop severe metabolic alkalosis [1]. Congenital chloride diarrhea begins in fetal life, and previous case reports have described antenatal sonographic findings, including polyhydramnios and multiple distended bowel loops [1-7]. Although characteristic of congenital chloride diarrhea, these findings can also be observed in fetal small bowel mechanical obstruction, making diagnostic confluence [2]. Almost all published articles described the two main findings as: Polyhydramnios and multiple dilated loops - suspected ileal atresia. The postnatal clinical data revealed waterish diarrhea without meconium and the fluid mimicking urine colour and consistency [8].

RESULTS

The retrospective analysis discovers astonishing similarity between the dilated bowel loops and Turtle shell configuration. The dilated bowel loops appear uniformly distributed but not equal in size, what was striking is the polygonal fashion of the bowel loops suggestive of hypotonic bowels. In both siblings the postnatal abdominal radiographs show similar configuration of gas distended bowel loops with Turtle sign appearance. These findings are crucial for ruling out mechanical intestinal obstruction in neonates [9]. In both siblings the

MATERIALS AND METHODS

We present a case of 2 siblings, which antenatal ultrasound showed dilated bowel loops and Polyhydramnios. First sibling (Figure 1), Second siblings (Figure 2) in both siblings the initial reports described Polyhydramnios and multiple dilated loops - suspected ileal atresia. The postnatal clinical data revealed waterish diarrhea without meconium and the fluid is mimicking urine colour and consistency [8].

Figure 1: A. Antenatal US shows polyhydramnios
B, D, E. Antenatal US shows multiple uniformly dilated loops with polygonal structure
C. Turtle shell polygonal structure
F. Postnatal abdominal radiograph shows Turtle shell configuration of bowel loops.

Keywords:
Congenital chloride diarrhea; Functional bowel obstruction in neonates; Mechanical bowel obstruction in neonates
operations are avoided and prompt long term substitution of Sodium Chloride 5.85% and Potassium Chloride safe their life up to present.

CONCLUSION

We believe the Turtle sign is reliable sign for precise diagnosis of Congenital chloride Diarrhea. Obviously, there will be arguments that every case is different, but our strong opinion is: If the Turtle signs is seen it means functional - not mechanical obstruction. At least the antennal description of Turtle sign will prompt us for Differential diagnosis not for direct surgical treatment. Another potential significance: The Turtle sign probably will be seen in other types of functional obstruction. The future will clarify this idea.

REFERENCES


Figure 2: A. Antenatal US shows Polyhydramnios
B, D. Antenatal US shows multiple dilated bowel loops with polygonal structure
C. Antenatal 3D-US multiple dilated polygonal bowel loops
E. Turtle shell with polygonal structure
F. Postnatal abdominal radiograph shows polygonal gas distended loops