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Review Article

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

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ABSTRACT

Background: Congenital chloride diarrhea is a rare autosomal recessive disease characterized by profound watery diarrhea as a result of mutations in the gene encoding the intestinal chloride/bicarbonate exchanger. Congenital chloride diarrhea originates antenatally, and previous reports have described antenatal sonographic findings as, polyhydramnios and multiple distended bowel loops. But this is not enough. Dilated bowelscan be observed also in fetal mechanical bowels obstruction, which needs surgical attention to neonates. Our aim is to delineate some reliable sonographic sign of non mechanical bowel obstruction with goal - avoiding unnecessary operations.

Method: We compare the antenatal sonography of 2 siblings retrospectively and provide antenatal images as well abdominal postnatal radiographs.

Results: The antenatal sonographic images of both siblings show peculiar similarity which is not seen in mechanical bowel obstruction. All images show uniform distribution of dilated bowel loops with polygonal shape, which is sign of hypotonic bowels, not rounded dilated loops under pressure- typical for mechanical obstruction. The polygonal fashion of the bowels repeats the shape of turtle shell structure and so we label this feature as "Turtle sign".

Conclusion: We believe the Turtle sign is reliable signs for functional, not mechanical bowel obstruction. Such findings should prompt the clinicians for immediate conservative chloride substitution instead surgical treatment without definitive results. Once Turtle sign is recognized - the operation could be avoided

Keywords: Congenital chloridorhea; Functional bowel obstruction in neonates; Mechanical bowel obstruction in neonates

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 in utero 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 confuse [2]. Almost all published articles described the two main findings as: Polyhydramnios and multiple bowel loops dilatation. We found only one article [3] attempting more precise description of dilated loops describing them as "Honeycomb appearance "but usually the honeycomb cells are equal hexagonal structures.

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].

RESULTS

The retrospective analysis discovers astonish 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

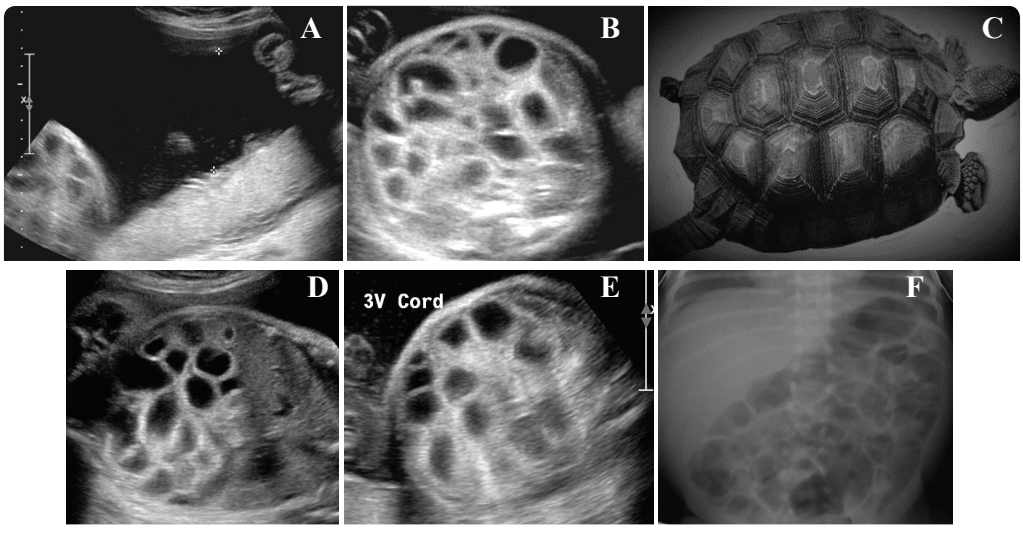

Figure 1: A. Antenatal US shows polyhydramnios
B, D, E. Antenatal US shows multiple uniformly dilated loops with polygonal structure
C. Turtle shell polygonal structure
F. Postnatal abdominal radiograph shows Turtle shell configuration of bowel loops.

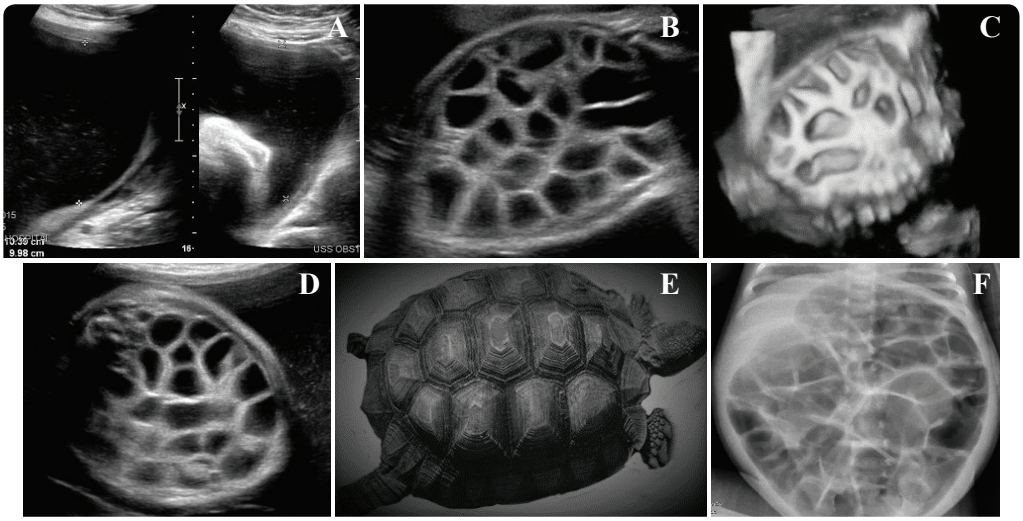

Figure 2: A. Antenala US shows Polyhydramnios
B, D. Antenatal US shows multiple dilated bowel loops with polyghonal structure
C. Antenatal 3D US-multiple dilated poligonal bowel loops
E. Turtle shell with polygonal structure
F. Postnatal abdominal radiographshows polygonal gas distended loops

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.

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