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DUODENAL ATRESIA: ANTENATAL DIAGNOSIS AND FUTURE

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INTRODUCTION :

Duodenal atresia is the most common cause of upper gastrointestinal obstruction that can be diagnosed antenatally. It is seen in 1 case out of 6,000 to 10,000 births [1] and it can be isolated or associated with other malformations. About 30% of infants with atresia have trisomy 21.

The diagnosis is prenatal in 80 to 90% of cases. Postnatally, it is generally evoked in the presence of vomiting, most often bilious, with or without anomalies of meconium elimination. The treatment is surgical without forgetting neonatal resuscitation which helps to improve the prognosis of this pathology, hence the interest of antenatal diagnosis for better neonatal care.

OBJECTIVE :

To clarify the contribution of imaging in the antenatal diagnosis and the future of duodenal atresia.

MATERIALS AND METHODS :

This is a descriptive retrospective study of the medical records of patients who had a prenatal diagnosis (ultrasound, MRI) of duodenal atresia, including five patients. The data is compiled from the archives of the Radiology B, Neonatology and Pediatric Surgery departments of Monastir over an extended period from 2010 to 2019. Initially, a number of nine cases were included in the study, then four were excluded due to incomplete medical records (imaging and/or operative reports not available) and one patient who delivered a stillborn (for lack of autopsy data).

RESULTS :

The incidence of duodenal atresia at CMNM is estimated at 1 case per 6350 live births and the average maternal age is 32 years with extremes ranging from 27 to 43 years. Consanguinity was noted in one case among the 5 cases. Multiparity was noted in two cases. One of the five pregnancies in our series was complicated by balanced gestational diabetes under diet.

The antenatal obstetric ultrasound allowed to suspect the diagnosis in the five cases with an average term of 28 weeks of amenorrhea (SA) and 6 days. In three cases the diagnosis was suspected on second trimester morphological ultrasound and in the other two cases the diagnosis was made during third trimester ultrasound. Ultrasound revealed a typical appearance with gastric and duodenal dilation creating the “double bubble” appearance, associated with an absence of visualization of the liquid in the downstream digestive loops, suggesting a complete duodenal obstacle (Figure 1). In two cases the downstream loops were visible containing fluid that could be explained either by an incomplete obstruction or by the late onset of the duodenal obstruction. The appearance of the coves downstream was not specified in two cases. In only one case, the digestive wall presented a thickened aspect with the presence of a layer of extra digestive liquid.

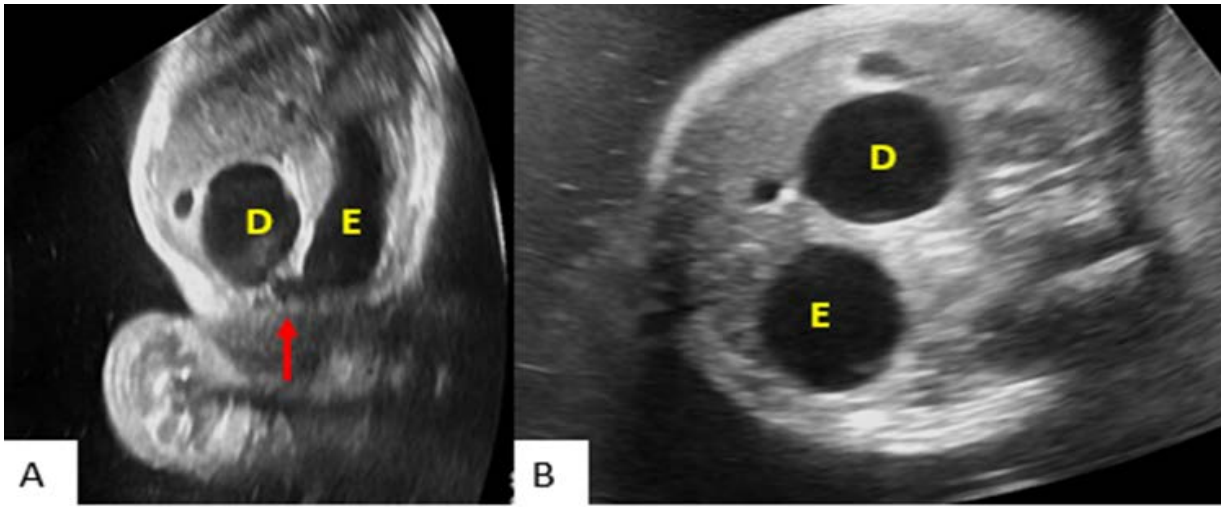

Figure 1 : Duodenal atresia in a 26 WA fetus: ultrasound sections of the axial (B) and parasagittal (A) fetal abdomen showing a typical appearance of a double water bubble (B) communicating (A, red arrow) (E: stomach, D : duodenum) with polyhydramnios

Fetal MRI was performed secondarily in two cases where the diagnosis of complete duodenal obstruction was doubtful: In the first case, the diagnosis was suspected on morphological ultrasound at 25HERin front of a double bubble aspect but with visibility of the digestive loops downstream suggesting an incomplete duodenal obstacle such as the duodenal diaphragm or annular pancreas. In the second case, the obstetrical ultrasound performed at 29 WA revealed distension of the digestive loops leading to the suspicion of a small bowel atresia-type malformation. In both cases the MRIfetus performed respectively at 37 WA and 31 WA, had shown severe distension of the stomach and duodenum with a tapered appearance of the distal end of the duodenal segment and Dflat downstream digestive loops in favor of a complete duodenal obstacle.(Figure 2).

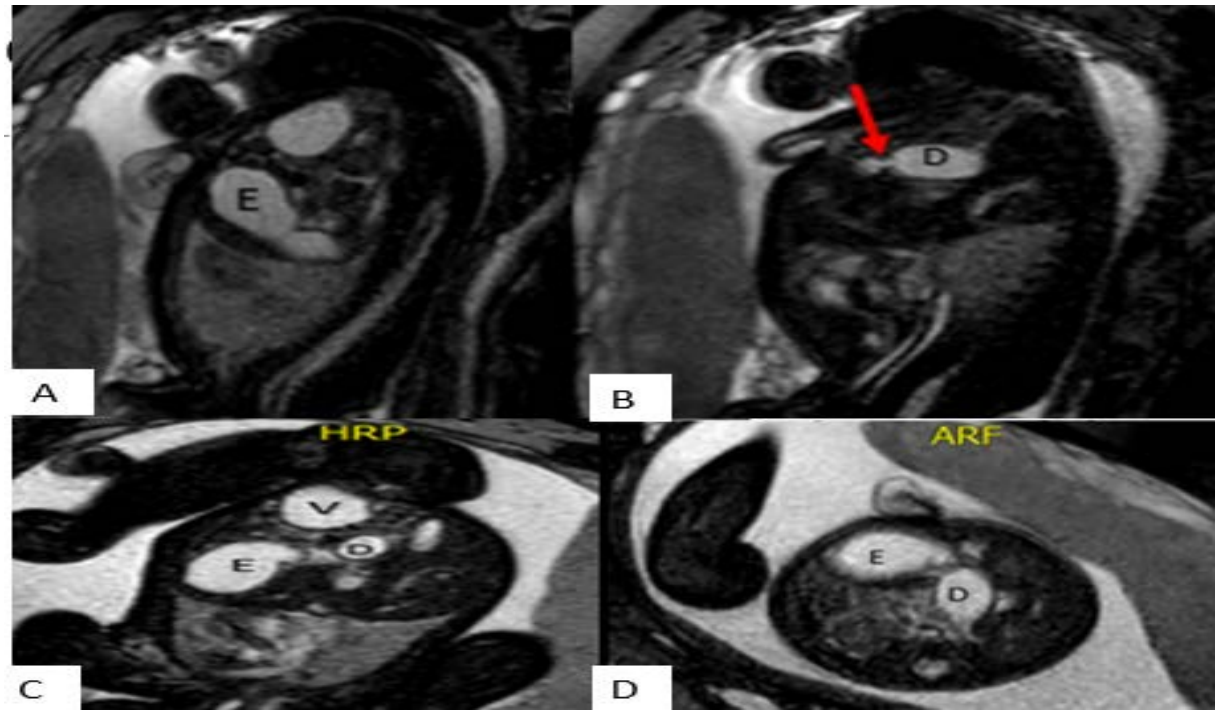

Figure 2: fetal MRI at 31 WA (A, B: sagittal T2, C: coronal T2, D: axial T2) showing gastro-duodenal distension (C, D) in a communicating "double bubble" (B: red arrow) with arrest in bird's beak of the duodenum without visualizationa fluid signal in the downstream digestive loops suggesting a complete duodenal obstruction(E: stomach, D: duodenum, V: bladder)

The average birth weight was 2850 g with extremities of 2200 g to 3750 g. There is a male predominance with a sex ratio of 4 boys for one girl. Three deliveries were made before 37 weeks of amenorrhea but not before 36 weeks of amenorrhea. Trisomy 21 was found in three of the five cases in our series. The annular pancreas in two cases. Apple Peel syndrome in 1 case. No other congenital malformations were found in particular no cardiac abnormality nor esophageal atresia. No amniocentesis was performed. A postnatal karyotype was requested in three cases in front of a birth examination showing facial dysmorphism which confirmed trisomy 21 for these three cases. Surgical intervention was performed in all cases on D1 of life by a latero-lateral duodeno-duodenostomy (5 cases). Surgical exploration showed duodenal atresia in five cases, associated with Apple Peel syndrome in one case and annular pancreas in two cases. The postoperative course was favorable in all cases, no complications were described.

DISCUSSION :

Duodenal atresia results from a recanalization defect; the bowel tube fails to channel its lumen which normally occurs during the sixth week of gestation. This lack of recanalization could explain certain forms of biliary atresia which can be associated with duodenal atresia. Depending on the nature of the obstacle, atresia is classified into three types (Figure 4): The vast majority (92%) are type I: an obstructing septum or web is formed by the mucosa with no defect in the musculature or mesentery (Fig.4A). Type II atresias, which represent only 1% of all duodenal atresias, consist of two blind ends of the duodenum connected by a short fibrous cord (Fig.4B). Type III atresia, with two blind ends of duodenum that are completely disconnected (Fig.4C), occurs in 7% of cases. Eighty-five percent of duodenal atresias are located at the junction of the first and second part of the duodenum.

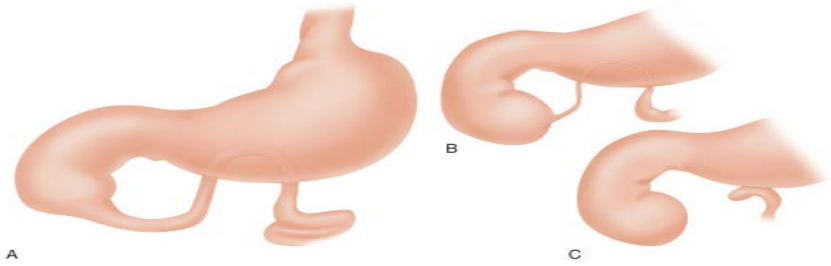

Figure 3: The different types of duodenal atresia [16]

4A: type I, 4B: type II, 4C: type III

The incidence of duodenal atresia is 1 in 6,000 to 10,000 births [1]. For the distribution of this pathology according to sex, according to what is described in the literature, we find a male predominance with a sex ratio of 2 to 3 [2], [4]. In the majority of reported series, the mean birth weight was 2 kg [4], [5]. For our series the average birth weight was 2850g which can be explained by births occurring between 36 and 37 SA. The antenatal diagnosis of duodenal atresia is, in the majority of cases, made by obstetric ultrasound. Indeed, the sensitivity of ultrasound varies from 87 to 94% of

cases depending on the study [6]. In most series the diagnosis is made from 20 WA during the second trimester ultrasound, few studies have evoked the diagnosis of duodenal atresia at a gestational age less than 20 WA [6,7]. From 29 SA, the amount of amniotic fluid swallowed by the fetus exceeds the resorption capacity of the gastroduodenal mucosa. This leads to abnormal dilation of the stomach and duodenum upstream of the obstacle causing polyhydramnios [3,4] which was consistent with our series where polyhydramnios was reported on ultrasound in all cases. In fact, this gastroduodenal distension produces the watery “double bubble” appearance which is a pathognomonic sign of duodenal obstruction. For our series, this sign was found in all the fetuses and on this sign the diagnosis of duodenal atresia was evoked. An upper transverse section of the abdomen reveals two well-limited fluid images, corresponding to the stomach and the initial part of the dilated duodenum. Faced with such an aspect, it is essential to visualize the continuity between the stomach and the duodenum, producing an hourglass image, which makes it possible to eliminate other differential diagnoses such as a bile duct cyst, a mesenteric cyst or a digestive duplication. [8].These two liquid images observed may contain fine echogenic particles (vernix or meconium) animated by movement under the effect of peristaltic waves of struggle. It is necessary to verify the permanence of these images during successive checks to be able to confirm the diagnosis. Indeed, one can observe transient images of gastric or even gastroduodenal dilation simply indicating a fetus vomiting or visualizing intestinal peristaltic movements. Fetal MRI is indicated as a second intention complementary to ultrasound when faced with a doubtful appearance. In fact, this technique allows, thanks to its excellent contrast resolution, to locate the seat of the anomaly more precisely than ultrasound. It is also superior in the precision of the nature of the obstruction since it visualizes a specific signal of the contents of the digestive tract allowing to make diagnoses more oriented on the complete or incomplete character of the duodenal obstacle as well as on the etiology. [7]. Indeed, the absence of liquid signal from the jejunal loops and the presence of a large colon and a discreetly reduced T1 signal is in favor of duodenal atresia. MRI can also be used to look for associated signs, in particular other obstructions in the context of multiple atresia. In our series, MRI was performed in two fetuses where the diagnosis was doubtful and it made it possible to evoke duodenal atresia which was confirmed intraoperatively in these two cases.

In 30 to 52% of cases, duodenal atresia is isolated [9]. However, it can be associated with trisomy 21 in 30% of cases, an annular pancreas in 23% of cases, congenital heart defects in 22% of cases, malrotation in 20% of cases, atresia esophagus in 8% of cases and other congenital malformations in 20% of cases [10].

In fact, the demonstration of a duodenal obstacle during an obstetric ultrasound and/or polyhydramnios requires an in-depth fetal assessment in search of chromosomal abnormalities and associated malformations, particularly digestive and cardiac.

Duodenal atresia is an infrequent malformation whose prognosis is generally preserved. Medical termination of pregnancy is not indicated once the diagnosis is suspected antenatally. However, once associated with other malformations, the antenatal karyotype must be performed. The vaginal route is the usual delivery route and recourse to caesarean section is not indicated to improve the prognosis of this malformation. This by emphasizing the need for delivery in a 3rd level maternity unit for better neonatal care.

Surgery is the only treatment for duodenal atresia [2].

The approach is a midline laparotomy with one of the following procedures:

- a latero-lateral duodeno-duodenostomy
- a gastro-jejunostomy
- a duodeno-jejunostomy

Currently, a new approach is increasingly used which is endoscopy. The laparoscope has the value of a small, micro-invasive incision and better recovery in the diagnosis and treatment of congenital duodenal atresia. Laparoscopic methods can be performed in neonates without risk and are appropriate for a term neonate with CO2 pneumoperitoneum tolerance [4,5].

CONCLUSION :

The pathology of the digestive tract in the fetus is rich and dominated by malformations whose prognosis can be reserved. The antenatal diagnosis of digestive abnormalities and in particular duodenal atresia makes it possible to program the reception and care of the newborn from birth. It has undergone major development over the past twenty years due to the progress made in the field of fetal imaging. This is based on ultrasound supplemented if necessary by fetal MRI. The prognosis is all the better if it is isolated and if its diagnosis is made antenatally, thus allowing better neonatal and surgical care. The poor prognostic factors remain mainly prematurity, low birth weight and association with other malformations.

The particularity of this pathology comes from the high frequency of the associated anomalies conditioning the prognosis of this malformation. Hence the interest of systematic antenatal ultrasound screening in the second trimester to recognize this malformation based on suggestive signs including the double bubble appearance which is pathognomonic for this duodenal obstruction. An analysis of the digestive loops downstream generally makes it possible to evoke a complete duodenal obstacle. However, this is not always obvious and sometimes requires the use of a fetal MRI which offers a better analysis and characterization of the type of this duodenal obstruction and to make a complete lesional assessment of the associated anomalies.

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