

AUTHORS' REPLY TO COMMENTS

REVIEWER COMMENTS:

Reviewer 1

Interesting case, should be published.

Author reply:

Thank you for your comment.

Reviewer 2

Some photos are diffuse and small, and this case presentation is still difficult to understand. The fine schemas for this patient's IAPF should be made for journal readers in the field of HPB physicians.

Author reply:

All figures were modified to obtain higher resolution and bigger icons accordingly to reviewer's comment. Figure 2 was simplified and made more comprehensible. To better explain the management of our case, an additional scheme of the patient's clinical course and of the therapeutic interventions was added in Figure 5.

Reviewer 3

This is an interesting case. It is well written and deserves the publication in this journal. Some minor comments are listed.

1. Did the authors have any endoscopic image to show direct evidence about source of gastrointestinal bleeding?

Author reply:

No, unfortunately it was impossible to recover any images of the procedure.

2. As mentioned by the authors, this child received diuretic treatment for refractory ascites. What is the dosage of diuretics? How about the results of ascites?

Author reply:

At presentation, the infant had severe ascites for which administration of furosemide 1 mg/kg/die (divided in two doses) and spironolactone 2 mg/kg/die (divided in two doses) was started. Despite the maximization of the diuretic treatment, the ascites didn't improve and the patient presented acute respiratory distress due to abdominal distension. Therefore, daily paracentesis was required to improve the clinical status of the child. Paracentesis were carried out by the insertion of a percutaneous pigtail abdominal drainage, which permitted to improve the clinical status of the patient and to perform an accurate fluid balance between out-put (ascites, diuresis) and in-put (fluids, drugs) in order to maintain the patient's hemodynamic stability. The ascites improved only after the combined endovascular-surgical procedure, thus paracentesis were not further required (the abdominal pigtail was removed) and the diuretic treatment was gradually suspended. Since 3 months after the procedure, the patient has been free of diuretic treatment and has not presented ascites anymore. These data have been added in the revisited manuscript.

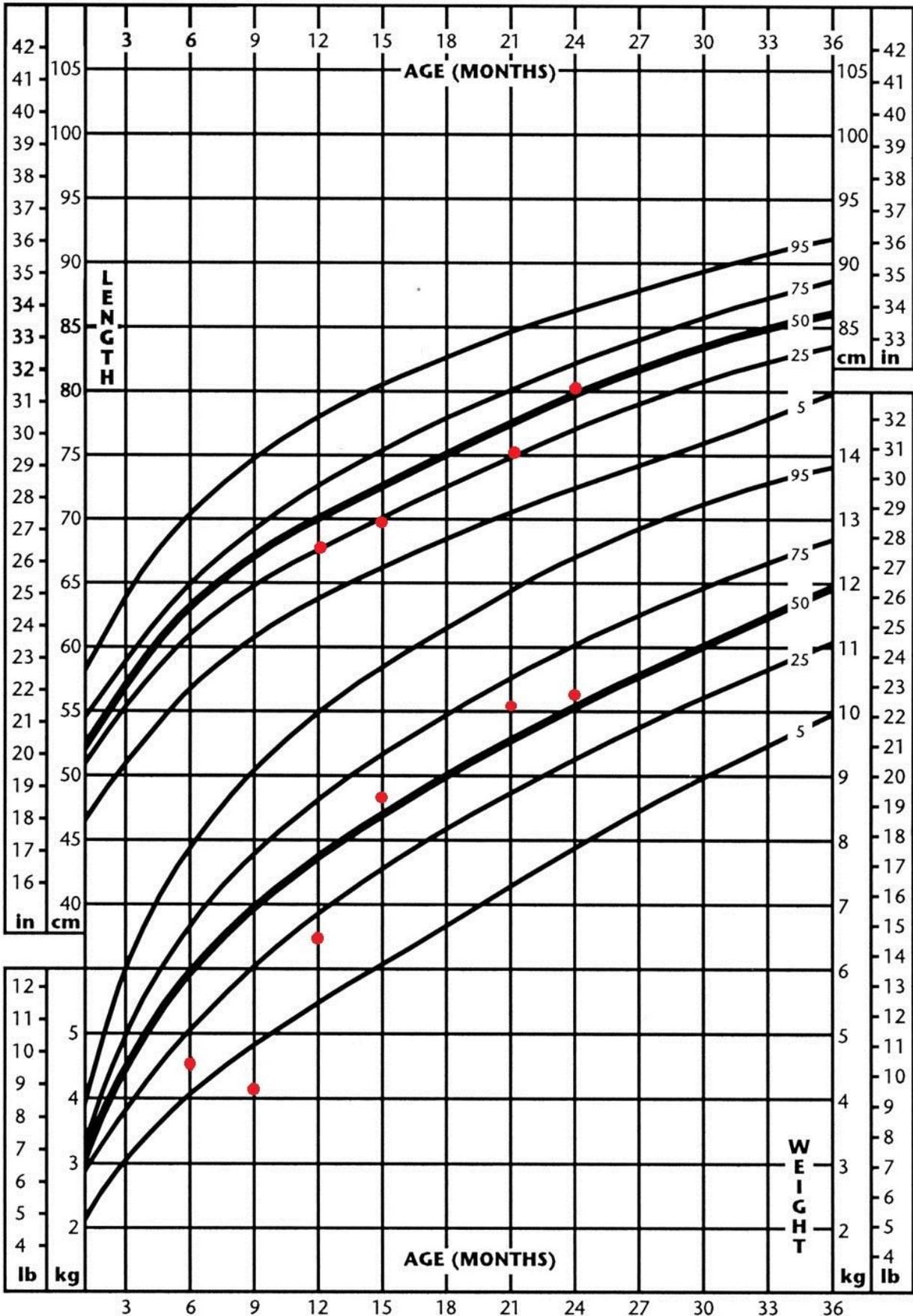
3. How about this child's growth?

Author reply:

The child was born weighting 2.6 kg. At 6 months of age, when she presented to our unit, the patient was 4.5 kg (<25°) and during the following 3 months the child loss 500 g (<5°) despite an adequate caloric intake. After the complete occlusion of the IAPF by the hybrid endovascular-surgical procedure, the child rapidly improved her growth, reaching a weight of 8.66 kg (>50°) and height of 47 cm (25°) at 15 months. At 24 months of age (last follow-up) the patient as an appropriate growth [weight: 10.2 kg (>50°); height: 80 cm (>50°)] according to the growth curve for female affected by Down Syndrome (patient's physical growth curve attached). As the resolution of the IAPF by the combined endovascular-surgical procedure had a beneficial effect also on the child's growth, we included these information in the revisited manuscript.

**DOWN SYNDROME GIRLS
PHYSICAL GROWTH:
1 TO 36 MONTHS**

NAME _____ RECORD # _____



4. *When the authors said "... to avoid further invasive surgery", please tell us what invasive surgery is.*

Author reply:

Thank you for your question. As “further invasive surgery”, we considered major surgical procedures as liver resection, surgical shunting or liver transplantation. According to the current literature, IAPF not resolved by radiological embolization treatment, often require major liver resection, porto-caval shunt or liver transplantation, which might represent aggressive surgery for small infants and are associated with morbidity and mortality. Therefore, these surgical procedures should be left as last options in the algorithm treatment of congenital IAPF. The sentence “further invasive surgery” was rephrased in order to be more detailed.

5. *Language should be carefully improved, such as "...) and complete obliterated the fistula and restored normal PV hemodynamics ...", "... fed by major arterials ..."*

Author reply:

As documented by the language editing certificate, the manuscript had been edited by a recognized language editing services before the initial submission. An additional language editing by native English-speaking was performed for the revisited version of the manuscript.