

The malrotated baby – a diagnostic dilemma

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ABSTRACT

We present a case in which misinterpretation of radiological images resulted in an incorrect diagnosis of malrotation. This case highlights the importance of adequate radiological review of images and of ensuring that the clinical picture matches the findings. Had this not been questioned in this case the baby could have undergone an unnecessary operation with potential mortality and morbidity.

CASE REPORT

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A 3 week old, premature baby girl, born at 30 weeks (twin II) was noted to be suffering from intermittent, spontaneously resolving abdominal distension which had been present from birth. Following the introduction of full feeds this worsened and she had a single episode of pale green vomit on day 25 of life. A diagnosis of malrotation was suspected and confirmed on upper gastrointestinal study (Fig. 1). Conservative management was instituted and she was transferred the same day to a tertiary centre for further management.

On arrival she was noted to be clinically well, except for a mildly distended abdomen and irregular bowel motions. History of vomiting was not confirmed. In view of such an atypical clinical presentation for malrotation the external images were reviewed. It was noted from anatomical landmarks that the film was rotated, with two thirds of the heart lying to the right of the spine, and only a third to the left. A repeat upper GI contrast study confirmed normal bowel alignment and filling (Fig. 2). The heart shadow is normally orientated and the duodenojejunal junction is seen to the left of a vertebral body pedicle and at the level of the pylorus, confirming normal alignment. A rectal biopsy excluded Hirschsprung and thus the working diagnosis was that of

immature gut motility. The patient was recommenced on feeds and is well at 3 months of actual age.

DISCUSSION

During normal embryological development, the midgut rotates counterclockwise through 270 degrees around an axis defined by the superior mesenteric artery, to lie in its final anatomical position. Malrotation occurs in approximately 1 in 500 births and refers to a range of failures of this normal embryological development, with absent or incomplete rotation and fixation. Malrotation can present with a variety of symptoms, varying from acute to chronic and generally within the first year of life, although it can present at any age. 75% of symptomatic cases occur in newborns, but this condition can remain asymptomatic throughout life or present with atypical clinical findings. The classical clinical manifestation in the neonatal group is of bilious vomiting with abdominal distension.

Volvulus of the mid-intestine is a life-threatening complication of intestinal malrotation with historical mortality rates as high as 23% (1). Hence accurate diagnosis and

subsequent surgical management is crucial. Upper gastrointestinal contrast study is currently the imaging modality of choice, with quoted sensitivity of 93-100% (2). Normal positioned bowel and malrotation are distinguished by identification of the duodenojejunal junction, the normal position lying to the left of the left-sided pedicles of the vertebral body at the level of the duodenal bulb.

However, the diagnosis of malrotation on the basis of an upper GI series can be challenging, even to an experienced radiologist. A 15% false-positive rate was reported in two studies looking at the detection of malrotation in children and infants who subsequently underwent surgery (3). Similarly false-negative rates of 3-6% have also been described. Thus meticulous technique and reporting is clearly essential, with appreciation of normal variants and recognition of anatomic subtleties. This case illustrates the importance both of adequate radiological review of images and of ensuring that the clinical picture matches the findings. Had this not been questioned in this case the baby could have undergone an unnecessary operation with potential mortality and morbidity.

TEACHING POINT

The diagnosis of malrotation can be challenging and relies on thorough clinical and radiological evaluations. It is essential to ensure adequate radiological images are obtained and that these correspond to the clinical picture.

ABBREVIATIONS

GI = Gastrointestinal

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KEYWORDS

Malrotation, radiological diagnosis, UGI, upper GI study

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FIGURES

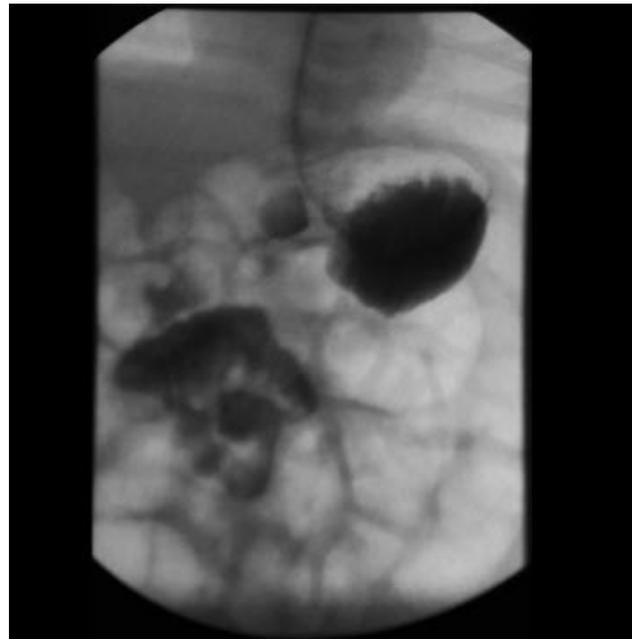


Figure 1: 3 week old baby girl with initially misdiagnosed malrotation due to patient rotation. Initial upper gastrointestinal contrast study suggesting a diagnosis of malrotation. On closer examination it can be seen that the film is rotated, with two thirds of the heart lying to the right of the spine, and only a third to the left.

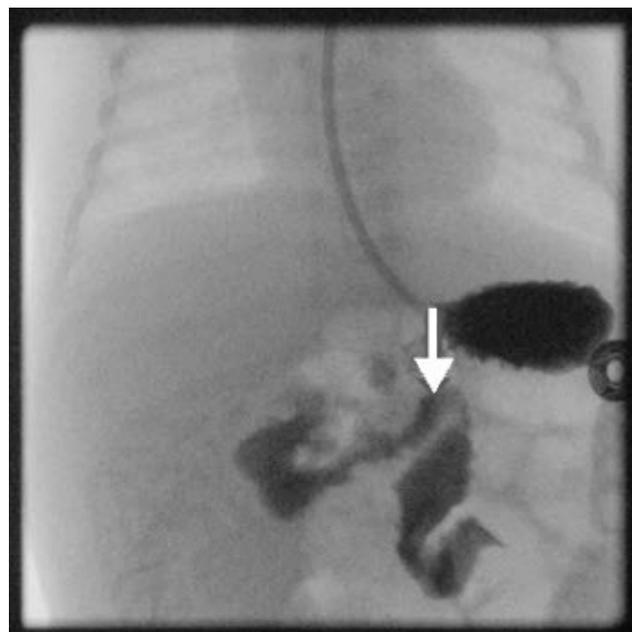


Figure 2: 3 week old baby girl with initially misdiagnosed malrotation due to patient rotation. A repeat upper gastrointestinal contrast study demonstrating normal bowel alignment and filling. The film is well centred with only one third of the heart lying to the right of the spine, and two thirds to the left. The duodenojejunal junction (arrow) is seen to the left of a vertebral body pedicle and at the level of the pylorus, confirming normal alignment.