



Case reports

# Right-sided diaphragmatic hernia in infants after liver transplantation

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**Abstract** Liver transplantation is just as successful in infants as in older children, but more challenging. This relates to the low weight of the recipients and to their rapidly deteriorating clinical condition (malnutrition and end-stage liver disease) (*J Pediatr* 1990;117:205-210; *BMJ* 1993;307:825-828; *Ann Surg* 1996;223:658-664; *Transplantation* 1997;64:242-248; *J Pediatr Surg* 1998;33:20-23). In addition, higher rates of diaphragmatic complications have been shown to significantly correlate with a younger age (*Transplantation* 2002;73:228-232; *Transpl Int* 1998;11:281-283; *Pediatr Transplant* 2000;4:39-44), but diaphragmatic hernia has never been reported as a complication of liver transplantation. In this report, 2 patients who developed diaphragmatic hernia after liver transplantation are presented. The possible role of several contributing factors resulting in diaphragmatic hernia is discussed. These factors include (1) diaphragm thinness related to low weight and malnutrition, (2) direct trauma at operation (dissection and diathermy), (3) increased abdominal pressure after transplantation caused by the use of a slightly oversized liver graft, and (4) the medial positioning of the partial liver graft in the abdomen.

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Liver transplantation has developed over time in children of all age groups, and it has been shown that results in infants are satisfactory [1-5]. However, the need for transplantation in infancy implies that the liver disease is rapidly progressing. As a result, infants who are candidates for transplantation are usually in a poor clinical state with a combination of malnutrition and complications related to an end-stage liver disease. The association of young age and very low weight makes transplantation a challenging intervention, and higher morbidity rates have been reported [1-3,6].

Younger patients are therefore at risk for higher rates of diaphragmatic complication after pediatric liver transplan-

tation [7-9], but diaphragmatic hernia has not been reported as a unique occurrence.

In this report, the authors outline 2 cases of diaphragmatic hernia that are complicating liver transplantation and discuss the possible contributing factors.

## 1. Case reports

Table 1 shows the details of 2 patients and gives relevant information regarding the transplant operation and the perioperative care. Patient 1 had Alagille syndrome, and patient 2 had biliary atresia and had an unsuccessful Kasai portoenterostomy operation. Both were critically ill and were hospitalized while awaiting transplantation because of complications related to end-stage liver disease. They both

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**Table 1** Peritransplant characteristics and evolution in patients 1 and 2

	Patient 1	Patient 2
<i>Patient</i>		
Sex	Female	Female
Primary disease	Alagille syndrome	Biliary atresia
Associated disease	Pulmonary stenosis (peripheral) Renal impairment (hemofiltration)	–
Previous operation	–	Kasai
<i>Condition before transplant</i>		
Malnutrition	Severe	Severe
Hospitalized	Yes (intensive therapy unit)	Yes
Ascitic decompensation	Yes (refractory)	Yes
<i>Transplantation</i>		
Age at transplant (mo)	9	5
Weight at transplant (kg)	5.3	5.8
Donor weight (kg)	64	30
Donor-recipient weight ratio	12	5
Graft type	Split <sup>a</sup>	Reduced <sup>a</sup>
Ischemic time (min)	833	844
Operative time (min)	450	327
Prosthetic abdominal closure	Yes	No
<i>Posttransplant</i>		
Time for extubation	13 d	27 h
ITU stay	15 d	38 h
Hemofiltration	Days 1 to 5	No
First week reoperation	N = 3	No

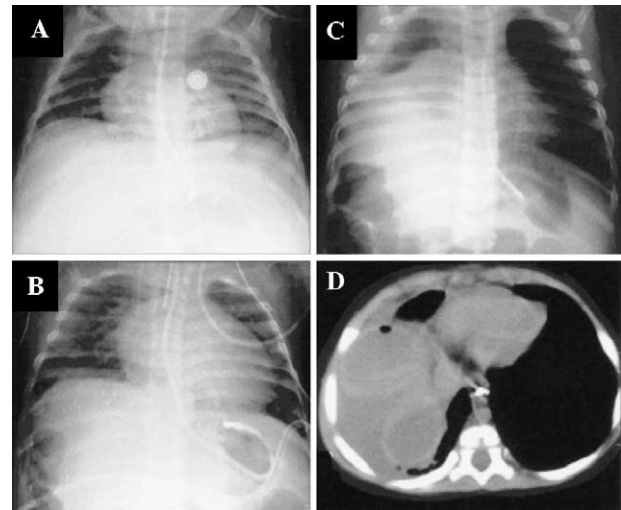
<sup>a</sup> Both grafts consisting of segments 2 and 3.

had severe malnutrition with *z* scores of  $-4.8$  and  $-3.9$  SD (patient 1), and  $-2.1$  and  $-1.9$  SD (patient 2) for weight and height, respectively, and all other anthropomorphic measurements being below  $-1$  SD (mid-arm circumference, triceps skin fold, and mid-arm muscle mass). Both were urgently allocated a liver graft from a larger donor while waiting in hospital (10 days in both cases). Patient 1 received a split liver graft and patient 2 received a reduced liver graft, both grafts consisting of segments 2 and 3 (left lateral segment graft according to American terminology).

The results of chest radiographs before the operation and before discharge from the hospital after transplantation were normal (Figs. 1A, B and 2A, B). The posttransplant history was as follows.

### 1.1. Patient 1

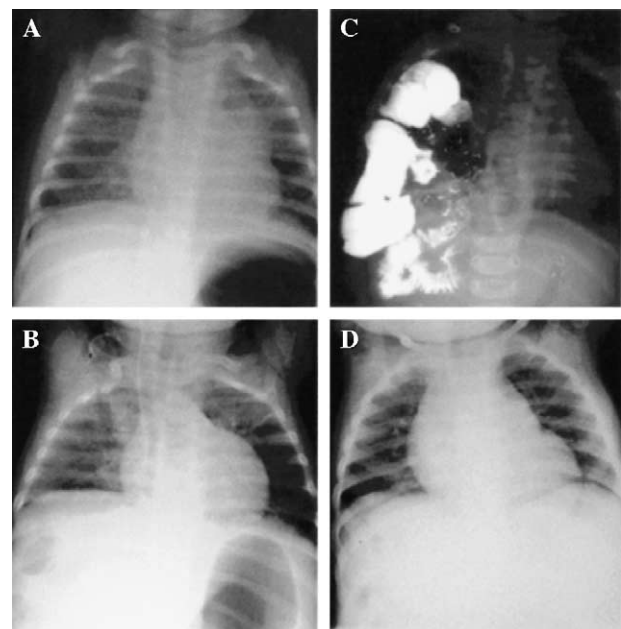
The posttransplant course was stormy, with the need for hemofiltration for 5 days and 3 reoperations during the first week (twice for hemorrhage and once for repositioning the liver graft to improve portal flow that had dampened).



**Fig. 1** Imaging in patient 1 at various times. A, Chest x-ray before transplant. B, Chest x-ray after transplant. C, Chest x-ray showing right-sided diaphragmatic hernia (10 months after transplant). D, Computed tomographic scan showing incarcerated bowel in the chest (10 months after transplant).

A further operation took place on day 28 to remove a SILASTIC sheet (Down Corning, Midland, Mich) used for a previous prosthetic abdominal closure [10], the latter procedure being necessary because of the use of an oversized liver graft for transplantation (large size mismatch with donor-recipient weight ratio = 12). She was discharged from the intensive care unit on day 15 and from the hospital on day 36. The first outpatient reviews were unremarkable.

Ten months later, she presented with respiratory distress. In the preceding week, she had some coryzal symptoms and



**Fig. 2** Chest x-rays in patient 2 at various times: 1 month of age (A), day 14 after transplant (5 months of age) (B), diaphragmatic hernia (9 months of age) (C), and follow-up (11 months of age) (D).

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