Biliary atresia is the most common indication for liver transplants in children, and this disorder is associated with some congenital vascular anomalies. Complex vascular anomalies increase the technical difficulties of the operation and previously resulted in high mortality in these patients. The role of the radiologist is to define the conditions in which transplantation is high risk or contraindicated and to identify any anatomic variations that may alter the surgical approach.\(^1\) The purpose of this study is to define the congenital vascular abnormal conditions in biliary atresia patients that would alter the surgical procedure during transplantation.

**MATERIALS AND METHODS**

During a 7-year period, from December 1993 to November 2000, 73 potential pediatric recipients due to biliary atresia received pretransplant evaluation. They were aged 3 months to 12 years 5 months (mean 26.59 ± 24.82 months) with a body weight of 5.2 to 36.1 kg (mean 11.27 ± 5.05 kg). These potential candidates received catheter angiography as a part of a pretransplantation survey. Angiographies were performed using digital subtraction angiography techniques with the femoral artery approach. Under general anesthesia, the visceral catheter was placed in the abdominal aorta and common hepatic artery (CHA) from celiac axis RHA from SMA (n = 5); from inferior phrenic artery (n = 1), and from right renal artery (n = 1). Portal vein and IVC anomalies included: (8) engorged retroperitoneal varices (n = 33), including spleno-renal shunt (n = 4) and porto-hemiazygos shunt (n = 2); (9) MPV thrombosis (n = 10); (10) small portal vein <4 mm in caliber; (11) hepatofugal flow of PV (n = 10); (12) interrupted IVC (n = 1) and small IVC with prominent azygos and hemiazygos veins (n = 1). The groups (4), (9), and (10), 27 cases (37.5%) in total, may need to alter major surgical procedure. Seven cases (9.7%), groups (7) and (12), are not good candidates for LRLT.

**RESULTS**

Catheter angiography failed in a small pediatric candidate (body weight 5.2 kg) due to technical difficulty puncturing the small access artery. The average diameter of common or proper hepatic artery (CHA or PHA) and main portal vein (MPV) were 0.35 ± 0.11 and 0.46 ± 0.24 cm, respectively. There were various major anatomic variations in hepatic arteries: (1) conventional anatomy (n = 46); (2) LHA from aorta and common hepatic artery (CHA) from celiac axis (n = 3); (3) CHA from celiac axis but LGA from PHA (n = 2); (4) replaced CHA from SMA (n = 7); (5) replaced RHA from SMA (n = 4); (6) common trunk of LHA and LGA from aorta (n = 5); (7) aberrant RHA (n = 5): from SMA with small CHA (n = 2), from aorta (n = 1), from inferior phrenic artery (n = 1), and from right renal artery (n = 1). Portal vein and IVC anomalies included: (8) engorged retroperitoneal varices (n = 33), including spleno-renal shunt (n = 4) and porto-hemiazygos shunt (n = 2); (9) MPV thrombosis (n = 10); (10) small portal vein <4 mm in caliber; (11) hepatofugal flow of PV (n = 10); (12) interrupted IVC (n = 1) and small IVC with prominent azygos and hemiazygos veins (n = 1). The groups (4), (9), and (10), 27 cases (37.5%) in total, may need to alter major surgical procedure. Seven cases (9.7%), groups (7) and (12), are not good candidates for LRLT.

**DISCUSSION**

Biliary atresia is the most common indication for liver transplants in children, and this disorder is associated with some congenital vascular anomalies in 10 to 27% of patients.\(^2\) Specific findings include: polysplenia, intestinal malrotation, absent prererenal inferior vena cava with azygous drainage, confluent hepatic veins to the right atrium, preduodenal portal vein, abnormal hepatic arterial supply, bilobed right lung, and situs inversus.\(^3\,4\) Many children with biliary atresia will progress to develop end-stage liver disease requiring definite treatment with a liver transplant, which can be technically complicated by the anatomic differences associated with this disorder. Some of these will preclude a normal surgery procedure; otherwise complications may occur. Vascular complications of the hepatic artery and portal vein are the major causes of graft failure in liver transplantation, especially in pediatric cases. The incidence of the hepatic artery thrombosis has been reported to range from 4% in adults to 12% in pediatric cases and up to 30% in children under 1 year of age, resulting in the most common cause of retransplantation.\(^5\,7\) This wide

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variation of values reflects the different means of diagnosis, characteristic of the vessels, vascular size, anomalies, site, and techniques of anastomosis.

In our series, 46 cases (63.9%) revealed conventional anatomy. They can receive either orthotopic liver transplantation or living related liver transplantation. Replaced CHA from SMA was found in seven cases (9.7%). That aberrant course of the artery deep to the portal vein may necessitate altering the sequence of the vascular anastomosis so the hepatic arterial reconstruction has to be performed prior to the portal vein anastomosis. An aortohepatic interposition graft is often necessary when adequate inflow cannot be ensured from the native hepatic artery, as in a small vessel or high-grade celiac axis stenosis. There were five cases (6.9%) that revealed aberrant RHA \( (n = 5) \) with small CHA: from SMA \( (n = 2) \), from aorta \( (n = 1) \), from right inferior phrenic artery \( (n = 1) \), and from right renal artery \( (n = 1) \). In this category, aortohepatic interposition or vascular grafts were required so they were excluded for LRLT in our series.

Engorged varices or with portosystemic shunt was found in 33 cases (45.8%) that need vascular ligation to maximize portal flow. Small portal vein \( (<4 \text{ mm}, 5 \text{ cases}) \) and portal vein thrombosis \( (10 \text{ cases}, 13.9\%) \) were troublesome in LRLT. Thrombectomy or vascular jump may be needed to solve this problem. Interrupted IVC or small caliber of IVC was another problem in liver transplantation, which may alter surgical anatomy.

CONCLUSION

Pathologic and normal variants of HA, PV, and IVC would alter surgical procedure as well as type of anastomosis or even graft interposition. Preoperative mapping of the vascular structure would guide the direction of surgical approach.

REFERENCES