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Case report

A case report of general anaesthesia for the surgeries of cleft lip-plate in an infant with congenital portosystemic venous shunt

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ABSTRACT

Congenital portosystemic venous shunt (CPSVS) is a rare complication in which the ductus venosus does not close naturally after birth and the shunt forms between the portal vein and vena cava, resulting in pathophysiological states, such as pulmonary hypertension, high serum galactose, hepatic encephalopathy and acute liver failure. Cleft palate is a more frequent congenital abnormality that can hamper feeding and physical growth. Both disorders present potential problems for surgical anaesthesia, including changes in hepatic drug metabolism and difficult airway management. This report summarises our experience of the managements of general anaesthesia for ear tube insertion and reconstructive surgery of cleft lip and cleft palate in a female infant with complication of CPSVS. A preoperative blood test showed high serum levels of aspartate aminotransferase and alanine aminotransferase but no clinically significant elongation of prothrombin time or activated partial thromboplastin time. The patient preoperatively received a strict galactose-free diet. Although general anaesthesia for two surgeries in this case were well managed and uneventful, appropriate preoperative galactose management and lower doses of medication are recommended even if preoperative liver function is normal in a CPSVS patient.

1. Introduction

Cleft lip and palate are corrected surgically in the first months of life if feeding is disrupted. However, the deformity needs not only sensitive surgical interventions but also careful anaesthetic management. Congenital porto-systemic venous shunt (CPSVS), a rare circulatory defect in which portal blood bypasses the liver, markedly impairing hepatic metabolism [1,2]. Thus, both conditions might require careful anaesthetic management. We report the case of anaesthesia for constructive surgeries of cleft lip and cleft palate lip in a female infant with complication of CPSVS.

2. Case report

The patient was a female born at 38 weeks and 5 days, weighing 3138 g. She was referred to our hospital's dental and oral surgery department from a nearby obstetrics and gynecology clinic after birth due to bilateral lip and jaw cleft palate. Guthrie screening test upon admission revealed hypergalactosaemia, elevated ammonia concentration and elevated bile acid levels. In further examinations, enhanced CT scan showed that the portal vein and inferior vena cava (IVC) were directly connected after merger of the superior mesenteric vein (SMV) and splenic vein (SPV), diagnosing congenital portosystemic venous shunt (Fig. 1). As external deformities such as enlarged fontanels were also observed, a hereditary disease was suspected, but the guardian refused to pursue chromosome analysis. The paediatric department began administering galactose-free milk for the treatment of hypergalactosaemia. The infant exhibited brachycephalia, but no intracranial compression was indicated to keep her under the follow-up observation.

Initial reconstructive surgery was performed on the cleft lips. It is common to perform the first lip reconstructive surgery for cleft

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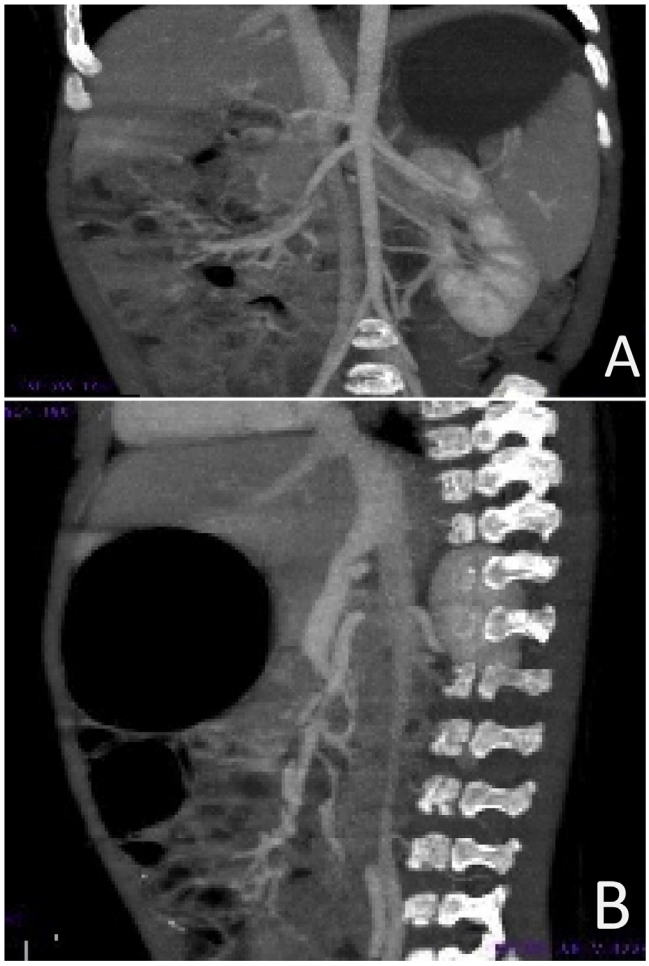

Fig. 1. A B: The portal vein and inferior vena cava (IVC) were directly connected after merger of the superior mesenteric vein (SMV) and splenic vein (SPV).

Table 1
Blood test data of hepatic system pre- and post- operation (1st and 2nd Operation).

	1st		2nd		
	Pre	Post	Pre	Post	NORMAL VALUES
Galactose	0.3		0.3	0.5	<3 mg/dL
Ammonia	65	39	77	45	12–66 μg/dL
Total Bile Acid	161.4		306.3	128.5	<10.0 μmol/L
AST	142	166	74	70	13–33 U/L
ALT	87	79	26	33	8–42 U/L

lip and palate three to six months after birth, but since this patient had difficult nursing through the mouth and her weight gain was small, surgery was delayed until month nine (at that time, height was 68.5 cm and weight was 7.4 kg). It is said that the functionality of the Eustachian tube is generally worse in children with cleft lips and cleft palates, making it easier for them to suffer from otitis media with effusion. Thus, ear tube surgery for an otitis media infection was simultaneously scheduled. A preoperative blood test showed high levels of aspartate aminotransferase (AST) and alanine aminotransferase (ALT) (Table 1), but no clinically significant elongation of prothrombin time (PT) or activated partial thromboplastin time (APTT) (Table 2).

Preoperative medication was not administered. General anaesthesia was gradually inducted using 3-L/min nitrous oxide, 2-L/min oxygen and sevoflurane. After the patient went to sleep, intravenous line for fluid and drug administration was kept using a 22G catheter in the left hand. She was administered 20-μg fentanyl citrate and 10-mg rocuronium bromide as a muscle relaxant, followed by tracheal intubation with a 4.0-mm diameter Ring, Adair and Elwyn (RAE) tube (without cuff) (Japan Medicalnext Co., Ltd, Osaka, Japan) without any difficulty. Rocuronium was administrated to confirm train-of-four (TOF) counts of 0 using TOF watch (MSD Co., Ltd, Tokyo, Japan). Oxygen at 0.2–0.5 l/min, air at 0.62 l/min, sevoflurane at 2%–3% and remifentanil at 0.08–0.1 μg/kg/min were administered during the surgery. Infiltration anaesthesia consisting of lidocaine containing 1/80,000 adrenalin was locally applied at the wound. Before starting the operation, 100-mg acetaminophen suppository was administered for post-operative pain. During surgery, the EKG showed no significant change, and oxyhaemoglobin saturation measured by pulse oximetry (SpO2) was maintained at 98%–100%. Upon completion of the surgery, anaesthesia was discontinued. Six minutes after the end of the procedure, the patient resumed spontaneous breathing. The patient was extubated 20 min after surgery upon confirming her sufficient respiratory status. Recovery of the TOF ratio to 100% was confirmed without the reversal agent for rocuronium. The operation and anaesthesia durations were three hours and thirty minutes and four hours and forty-four minutes, respectively. During anaesthesia, modified (glucose added) acetate Ringer solution was administered at the flow rate of 63 ml/h, and the total volume was 300 ml. There were no problems in controlling bleeding during surgery and no delay in recovery from anaesthesia.

Table 2
Blood test data of PT and APTT pre- and post-operation (1st and 2nd operations).

	1st		2nd		
	Pre	Post	Pre	Post	NORMAL VALUES
PT (s)	17.3	14.4	18.0	16.1	10.6–14.9 (s)
PT (%)	42.6	61.6	41.1	51.1	70–130 (%)
PT INR	1.46	1.23	1.53	1.37	0.87–1.2
APTT	52.5	32.9	39.2	47.6	23.3–38.2 (s)

Following the first operation, the second surgery for correction on the portion of the nostril was performed at 1.7 years of age, at which time patient height was 75.9 cm and weight was 8.5 kg. Preoperative blood tests still showed elevated AST and ALT (Table 1) and no elongation of PT or APTT (Table 2). Because there were no major problems with the first operation, we decided to use the same anaesthetic agents for anaesthesia.

Preoperative medication was not administered. For anaesthesia induction, 4-L/min nitrous oxide, 4–5-L/min oxygen and 3.5%–8% sevoflurane were gradually inhaled via mask. After the patient went to sleep, intravenous administration was started with a 22G catheter in the left hand for administration of 20-μg fentanyl, 10-mg rocuronium bromide and 0.02–0.07 μg/kg/min remifentanil. Rocuronium was administrated to keep TOF counts of 0. Once muscle relaxation occurred, the patient was intubated with a 4.0-mm diameter RAE tube, which was fixed in the midline of the lower jaw at a depth of 13 cm from the oral edge. Recovery of the TOF ratio to 100% was attained with 2 mg/kg of sugammadex.The operation and anaesthesia durations for the second surgery were two hours and thirty minutes and three hours and forty-three minutes, respectively. During anaesthesia, modified (glucose added) acetate Ringer solution was administered at the flow rate of 73 ml/h, and total volume was 270 ml. The course of this second anaesthesia was uneventful.

3. Discussion

Congenital porto-systemic venous shunt (CPSVS) is a rare circulatory defect in which portal blood bypasses the liver. The overall incidence of congenital portosystemic shunt is close to 1:30000 births [3,4]. Raskin et al. suggested that CPSVS occurs in the right

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