



Anesthesia for Cardiac Surgical Procedures in Alagille's Patients: an Institutional Experience

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Background

Alagille syndrome is a rare genetic disorder that can have cardiac and/or liver manifestations.

Peripheral pulmonary stenosis occurs in 90% of these patients thus requiring open-heart surgery and cardiopulmonary bypass.

The paucity of intrahepatic bile ducts causes hepatic dysfunction sometimes necessitating liver transplantation.

As a tertiary referral center for these patients, we describe our anesthetic experience.

Methods

After IRB approval, a retrospective chart review (2004-2017) was performed in our total series of 130 Alagille patients undergoing 104 cardiac surgical procedures at Lucile Packard Children's Hospital/Stanford University.

References

Bauser-Heaton H , McElhinney D, Hanley F, Wise-Faberowski L et al. *Circ Cardiovasc Interv* 2017.

Yildiz Ts, Yumuk NO, Baykal D et al. *Paediatr Anaesth* 2007.

Results

The majority (70/104) of patients undergoing cardiac surgery procedures were male. 11.9% were born prematurely. The mean age was 3.4 +/- 1.6 years and the mean weight 13.8 +/- 12.7 kg.

The majority of cardiac surgery procedures (46%) were unifocalization/pulmonary artery reconstruction, 34.8% were initial repairs and 65.2% being redo sternotomy. All cardiac surgery procedures required cardiopulmonary bypass except for 1 (coarctation of the aorta).

As expected, the majority of patients had a greater defect in biliary excretion rather than hepatic metabolism and synthesis (**Table 1**).

Most patients (68.4%) underwent an IV induction with fentanyl (89.5%) and another agent (ketamine 42% or versed 31.6%). All patients were maintained with fentanyl (62.3 +/- 72.6 mcg/kg/case) and another agent (volatile 47.4%, ketamine 42% or versed 63.2%).

Anesthetic duration was 616.35 +/- 162 min with average NIRS values 65.3 +/- 14.8%. All patients had a NIRS value less than 80% and 75% had a NIRS value < 50%. Surgical duration was 449.35 +/- 158.38 min with bypass times of 243.64 +/- 120.36 min and cross-clamp times of 35.67 +/- 18.05 min.

Transfusion was massive (92 +/- 38 cc/kg blood products) (**Table 2**) and 22 patients received FEIBA.

Most patients arrived to the CVICU on a fentanyl infusion and another agent and were ventilated 107 +/- 106 hrs.

There were 6 deaths, 0 within 48hrs and 1 within 30 days. There were 4 complications: 3 ECMO and 1 infection.

Table 1: Pre-operative data for Alagille's Patients

Baseline labs	Mean +/- SD
Platelet count	256 +/-103 K/uL
INR	1.15 +/- 0.07 sec
AST	87.5 +/- 63.6 U/L
ALT	76.5 +/- 59 U/L
Total bilirubin	3.1+/-5.4 mg/dL
GGT	223.3 +/- 200 U/L

Table 2: Intra-operative data for Alagille's Patients

	Duration	Mean +/- SD
Anesthesia		616.35 +/- 162 min
Surgical		449.35 +/- 158.35 min
Cardiopulmonary bypass		243.64 +/- 120.36 min
Cross clamp		35.67 +/- 18.05 min
	Intra-operative Transfusion	Mean +/- SD
PRBC		35.8 +/- 25.2 cc/kg
FFP		36+/- 26.1 cc/kg
CRYO		5.6 +/- 3.3 cc/kg
PLTs		14.4 +/- 13.4 cc/kg

Discussion

This is the first case series of Alagille patients undergoing cardiac surgery.

There were no anesthetic complications. Surgical procedures were long and complications, especially mortality (~6%), were higher when compared to similar surgical procedures in non-Alagille patients (1.6%).