

A two month male baby with Alagille Syndrome, posted for pyeloplasty: Anaesthesia management

Sir,

A 2-month-old male baby weighing 4 kg was brought to the hospital with a history of fever for 2 days and jaundice for one week. He was born to non-consanguineous parents and was delivered by emergency caesarean section in view of meconium-stained liquor. The baby had physiological jaundice and respiratory distress at birth and was treated in the neonatal intensive care unit (NICU) before discharge. The child was immunised appropriately.

The relevant laboratory reports were: total leucocyte count 16900 cells/cu mm, total bilirubin 14 mg/dL (direct 5 mg/dL), serum glutamic oxaloacetic transaminase (SGOT) 649 U/L, alkaline phosphatase (ALP) 438 IU/L, activated partial thromboplastin time (aPTT) 41.7 s. Ultrasonography (USG) of abdominal-pelvis showed gross right hydro-uretero-nephrosis with cortical thinning and mild hepato-splenomegaly. Tc ethylene dicysteine scan showed right pelvic-ureteric junction obstruction. Tc 99 mebrofenin hepatobiliary scan showed enlarged liver with impaired hepatocyte function. Mildly dilated right atrium (RA) and right ventricle (RV), moderate pulmonary branch stenosis predominantly at the left pulmonary artery (LPA)

with a gradient of 40 mmHg, a small patent foramen ovale (PFO) with the left to right shunt and mild tricuspid regurgitation were seen on echocardiography.

The child was stabilised with vitamin K, fresh frozen plasma (FFP) and antibiotics. A diagnosis of Alagille syndrome with obstructive uropathy and unconjugated hyperbilirubinemia was made, and the child was scheduled for right pyeloplasty. Relevant findings on pre-operative assessment were prominent forehead and pointed chin with normal temporomandibular joint mobility.

The standard monitoring-electrocardiography (ECG), oxygen saturation (SpO₂), non-invasive blood pressure (NIBP), precordial stethoscope, temperature and capnography was started in the operating room. The baseline vitals were stable. A 22-G intravenous cannula was secured in the right upper limb. 100% oxygen was given by facemask. A radiant warmer with a preset temperature of 37°C was used. The child was premedicated with 0.4 mg of ondansetron, 0.04 mg of glycopyrrolate, 0.1 mg of midazolam and 8 mcg of fentanyl intravenously.

General anaesthesia was induced with 10 mg of propofol, and 5 mg of succinylcholine was used to facilitate tracheal intubation (uncuffed size 3 endotracheal tube). As the pre-operative values of platelet count and prothrombin time-international normalised ratio (PT-INR) were normal, caudal block was given with 2.5 mL of 0.25% bupivacaine. Anaesthesia was maintained with sevoflurane in oxygen-nitrous oxide, and atracurium (2 mg initially and 0.5 mg top up) was used for muscle relaxation.



Figure 1: 2 month old baby with Alagille syndrome showing triangular face and wide forehead

End-tidal CO₂ (EtCO₂) was maintained between 35 and 45 mmHg. Ringer lactate was given as per the Holliday Segar formula using an infusion pump.

Pyeloplasty was performed in 1 hour and 30 min. The patient remained haemodynamically stable and there were no intra-operative complications. The neuromuscular blockade was reversed with 0.04 mg of glycopyrrolate, 0.2 mg of neostigmine and the trachea extubated. A paracetamol suppository (120 mg) was inserted per rectum for post-operative analgesia and the child transferred to the post-operative room. Post-operative stay in the hospital was uneventful, and the child was discharged after a month.

Alagille syndrome (AGS) is an inherited autosomal dominant disorder in which jagged 1 gene mutation causes microdeletion of the twentieth chromosome. Its prevalence is estimated as 1 case per 70,000 individuals.^[1] It features five classical criteria: (1) intrahepatic cholestasis associated with ductopenia in liver biopsy (95%); (2) heart malformations, typically, stenosis or hypoplasia of the pulmonary artery (92%); (3) skeletal involvement, generally, butterfly vertebrae (70%); (4) eye anomalies, often posterior embryotoxon (78%); and (5) distinctive facial features (91%), such as wide forehead, ocular hypertelorism with sunken eyes, prominent ears, triangular face and straight nose.^[2-4] [Figure 1] Three out of these five criteria are required for the diagnosis, or two if there is a family history.^[5] Renal involvement is variable (19%–74%) while renal dysplasia is the predominant manifestation.^[4-6] Bilirubin, coagulation profile and vitamin K level should be checked pre-operatively. As hepatosplenomegaly encourages

regurgitation, a rapid sequence induction may be necessary to prevent aspiration. Caution should be exercised with drugs handled by the liver. Drugs that decrease hepatic blood flow (HBF) should be avoided. Maintain intravascular volume to preserve HBF. Regional anaesthesia plays a major role in post-operative pain relief in children and the classical techniques have an excellent safety record.^[7] Epidural anaesthesia may be preferred over opioids but coagulation status and vertebral anatomy should be checked. Caution should be exercised during transport and positioning as osteoporosis may be present.

Succinylcholine was used as difficulty in intubation was anticipated. Nitrous oxide was used to decrease the requirement of anaesthetic agents, and as the pulmonary arterial hypertension was moderate. Sevoflurane was preferred for the maintenance of anaesthesia because of its less myocardial depressant action. Atracurium was chosen as it is metabolised by plasma hydrolysis. We used the caudal block as the X-ray of the lumbosacral spine and coagulation were normal. The child tolerated the procedure well.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Deepa T, Shwetha Gopalaiah, Prabha Parthasarathy, Shreyavathi R

Department of Anaesthesia, Sathagiri Institute of Medical Sciences
And Research Centre, Bengaluru, Karnataka, India

Address for correspondence:

Dr. Deepa T,
15, Hesarghatta Main Road, Navy Layout, Chikkasandra,
Chikkabanavara, Bengaluru - 560 090, Karnataka, India.
E-mail: raagaranjani@gmail.com

Submitted: 06-Feb-2020

Revised: 18-Apr-2020

Accepted: 14-Jun-2020

Published: 31-Jul-2020

REFERENCES

1. Jacquet A, Guiochon-Mantel A, Noel LH, Sqalli T, Bedossa P, Hadchouel M, *et al.* Alagille syndrome in adult patients: It is never too late. *Am J Kidney Dis* 2007;49:705-9.
2. Turnpenny PL, Ellard S. Alagille syndrome: Pathogenesis, diagnosis and management. *Eur J Hum Genet* 2012;20:251-7.
3. Habib R, Dommergues JP, Gubler MC, Hadchouel M, Gautier M, Odievre M, *et al.* Glomerular mesangiolipidosis in Alagille syndrome. *Pediatr Nephrol* 1997;1:455-64.
4. Alagille D, Estrada A, Hadchouel M, Hadchouel M, Gautier M,

Odievre M, *et al.* Syndromic paucity of interlobular bile ducts (Alagille syndrome or arteriohepatic displasia): Review of 80 cases. *J Pediatr* 1987;110:195-200.

5. Quiros-Tejeira RE, Ament ME, Heyman MB, Martin MG, Rosenthal P, Hall TR, *et al.* Variable morbidity in Alagille syndrome: A review of 43 cases. *J Pediatr Gastroenterol Nutr* 1999;29:431-7.
6. Kamath BM, Podkameni G, Hutchinson AL, Leonard LD, Gerfen J, Krantz ID, *et al.* Renal anomalies in Alagille syndrome: A disease-defining feature. *Am J Med Genet A* 2012;158A: 85-9.
7. Ponde V. Recent trends in paediatric regional anaesthesia. *Indian J Anaesth* 2019;63:746-53.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/ija.IJA_102_20

How to cite this article: Deepa T, Gopalaiah S, Parthasarathy P, Shreyavathi R. A two month male baby with Alagille Syndrome, posted for pyeloplasty: Anaesthesia Management. *Indian J Anaesth* 2020;64:727-9.

© 2020 Indian Journal of Anaesthesia | Published by Wolters Kluwer - Medknow