Journal of Cardiothoracic and Vascular Anesthesia 31 (2017) 918–921

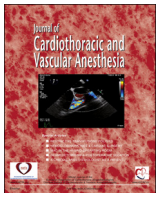
HOSTED BY

ELSEVIER

Contents lists available at ScienceDirect

ScienceDirect

journal homepage: www.jcvaonline.com



Twenty Years of Anesthetic and Perioperative Management of Patients With Tetralogy of Fallot With Absent Pulmonary Valve

John D. Jochman, MD[*], Douglas B. Atkinson, MD[†], Luis G. Quinonez, MD[‡], Morgan L. Brown, MD, PhD[†,1]

[*]*Department of Anesthesiology, University of Wisconsin, Madison, WI*
[†]*Department of Anesthesiology, Perioperative and Pain Medicine, Division of Cardiac Anesthesia, Boston Children's Hospital, Boston, MA*
[‡]*Division of Cardiovascular Surgery, Boston Children's Hospital, Boston, MA*

Objective: Review the authors' institutional experience of the induction and perioperative airway management of children with tetralogy of Fallot with an absent pulmonary valve.
Design: Retrospective chart review.
Setting: Large academic children's hospital.
Participants: Patients with the diagnosis of tetralogy of Fallot with absent pulmonary valve undergoing primary cardiac repair over a 20-year period.
Interventions: None.
Measurements and Main Results: Forty-four patients were identified with tetralogy of Fallot with an absent pulmonary valve from January 1995 through August 2014. Forty-two patients (95%) required surgery in their first year of life. Sixteen patients (36%) required mechanical ventilation preoperatively, including 11 neonates. Of the 28 patients not intubated preoperatively, only 1 (3.8%) exhibited minor airway obstruction following induction that was managed uneventfully. All intubations in the operating room were performed in the supine position. Five patients who were mechanically ventilated preoperatively in a lateral or prone position were supinated in the operating room without significant cardiopulmonary compromise. There were no patients who suffered cardiopulmonary arrest or required mechanical circulatory support. The median time to extubation was 2 days (range 1-13 days) in those patients who were not mechanically ventilated prior to their surgery. Ultimately, 5 patients required tracheostomy and 1 patient underwent lobectomy. Although, there was no mortality at 30 days, 4 children died within 1 year of their surgery. All the children who died had a genetic syndrome and required mechanical ventilation preoperatively.
Conclusions: While there are theoretical concerns specific to the induction and airway management of tetralogy of Fallot with an absent pulmonary valve, there were no episodes of cardiorespiratory arrest or extracorporeal membrane oxygenation in the authors' series. Neonatal age at the time of surgery, preoperative need for mechanical ventilation, and concomitant genetic syndromes are risk factors for respiratory morbidity. Mortality in this study was low compared to historic reports, likely reflecting improvement in surgical technique and intensive care management.

Key Words: congenital heart disease; airway; tetralogy of Fallot; absent pulmonary valve

[1]*Address reprint requests to* Morgan L. Brown, MD, PhD, Department of Anesthesiology, Perioperative and Pain Medicine, Cardiac Anesthesia Division, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115.
E-mail address: morgan.brown@childrens.harvard.edu (M. Brown).

http://dx.doi.org/10.1053/j.jvca.2017.02.006
1053-0770/

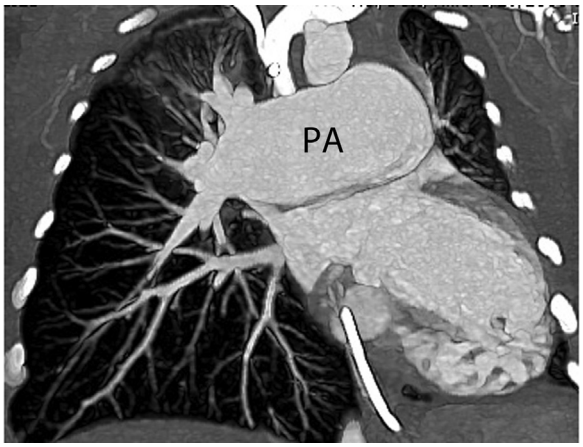


Fig 1. The enlargement of the pulmonary arteries causing compression of the bronchi in a neonate is visible on the chest CT scan. PA, pulmonary artery.

TETRALOGY OF FALLOT is a relatively common congenital cardiac lesion, with an estimated incidence of 421 cases per 1,000,000 live births.[1] Tetralogy of Fallot is defined by anterior deviation of the conal septum, with specific anatomic features including a conoventricular septal defect, overriding aorta, right ventricular outflow tract obstruction, and subsequent right ventricular hypertrophy. The spectrum of tetralogy of Fallot includes variations and degrees of severity. Tetralogy of Fallot with absent pulmonary valve is one such rare variant, estimated to occur in approximately 3% to 6% of all patients presenting with tetralogy of Fallot.[2]

A unique feature of tetralogy of Fallot with absent pulmonary valve is the development of marked aneurysmal dilation of the central and branch pulmonary arteries *in utero* resulting, in part, from increased flow and pulsatility related to the high pulmonary regurgitant fraction.[2] As a consequence of pulmonary artery enlargement, extrinsic compression of the trachea and bronchi often occurs leading to tracheomalacia and bronchomalacia (Fig 1). Smaller intraparenchymal airways also are obstructed by abnormal branching of segmental arteries.[3] These features result in a wide range of neonatal and infantile symptoms and clinical findings, including severe respiratory distress and cyanosis that may necessitate early surgical intervention. In the 1980s, the operative mortality reported for these patients was high (35-100%) due to pulmonary complications, although more recent series (2006 and 2007) have reported better early survival of 89% to 100%.[3–5]

Case reports have been published that discuss methods of anesthetic induction and airway management in patients with particularly severe respiratory compromise in the neonatal period. Hosking and Beynen have described a case in which an infant with tetralogy of Fallot with an absent pulmonary valve would suffer cyanotic spells whenever removed from a prone position and required an awake intubation in the semiprone position to maintain airway patency.[3] Similarly, at the authors' institution, Heinemann and Hanley reported a neonate who, even after intubation and mechanical ventilation, was intolerant of the supine position and needed an emergent sternotomy to relieve airway compression.[6] However, apart from these reports, there are few descriptions in the anesthesiology literature of the perioperative management of these patients. As tetralogy of Fallot with absent pulmonary valve may result in a spectrum of clinical presentations, the authors reviewed their institutional experience in caring for these patients, particularly as it pertains to problems encountered with anesthetic induction, airway management, and ventilation.

Methods

After obtaining Institutional Review Board approval, the authors conducted a retrospective chart review of all patients with the diagnosis of tetralogy of Fallot with absent pulmonary valve undergoing primary cardiac repair from January 1995 to August 2014. Information regarding patient age, sex, weight, cardiorespiratory symptoms, genetic disorders/anomalies, transthoracic echocardiography data, preoperative location/admission status, and need for respiratory support were recorded. Both electronic and paper anesthesia records were reviewed to identify information about anesthetic induction, airway management, and any occurrence of intraoperative airway obstruction or hemodynamic instability. Cardiac surgical management techniques, as well as ancillary airway procedures, were recorded, including rigid bronchoscopy, tracheostomy, tracheopexy, and lobectomy. Finally, information regarding postoperative outcomes and mortality was identified through chart review. Means with standard deviations, medians with minimum and maximum values, and incidences were calculated as appropriate based on the distribution and type of data. Univariate modeling using logistical regression and Kaplan-Meier survival curves were performed using JMP Pro 11.

Results

During the 20 years of review, 44 patients were identified with tetralogy of Fallot with an absent pulmonary valve who underwent primary surgery. Forty-two patients (95%) were less than 1 year of age at the time of the surgery. Patient demographics are detailed in Table 1. All patients presenting to surgery had echocardiographic evidence of severe pulmonary regurgitation, and nearly all (43 patients, 97.7%) had a preoperative diagnosis of aneurysmal pulmonary arteries. For the subgroup of 16 patients requiring preoperative mechanical ventilation for respiratory failure, 11 (69%) were neonates. All patients who were intubated preoperatively in the intensive care unit were intubated successfully in the supine position. However, 5 of these neonates required a non-supine position during mechanical ventilation to help alleviate airway obstruction preoperatively.

The majority of patients received an intravenous induction of anesthesia (71%) (Table 2). All patients received a muscle relaxant prior to endotracheal intubation, after confirming the ability to bag-mask ventilate with positive pressure. Of the 28 patients not intubated preoperatively, only one (3.8%) exhibited airway obstruction following induction. This was characterized by relatively high peak inspiratory pressures