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CASE SERIES

Bilateral Diaphragmatic Paralysis Following Paediatric Cardiac Surgery

Experience of four cases at the Royal Hospital, Muscat, Oman

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الشلل الحجابي ثنائي الجانب عقب جراحة قلبية في الأطفال

تجربة أربع حالات في المستشفى السلطاني، مسقط، عمان

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ABSTRACT: Bilateral diaphragmatic paralysis (BDP) is a rare complication of paediatric cardiac surgery. We report four children who developed BDP following cardiac surgery who were managed at the Royal Hospital, Muscat, Oman, between 2009 and 2014. All four children suffered severe respiratory distress soon after extubation and required re-intubation within two hours. In addition, all of the children underwent a tracheostomy as an interim method for ventilation. The four children were successfully weaned from positive pressure ventilation following the functional recovery of at least one side of the diaphragm.

Keywords: Diaphragmatic Paralysis; Heart Disease; Cardiac Surgical Procedures; Paediatrics; Case Series; Oman.

الملخص: يعد الشلل الحجابي ثنائي الجانب من المضاعفات النادرة في جراحة القلب عند الأطفال. ونستعرض هنا أربع حالات لأطفال أصيبوا بالشلل الحجابي ثنائي الجانب بعد جراحة في القلب، تم علاجهم في المستشفى السلطاني بمسقط في عمان بين عامي 2009 و 2014م. عانى الأطفال الأربعة من ضائقة تنفسية شديدة بمجرد نزع الأنبوب، وما كان مطلوبا من إعادة التنبيب خلال ساعتين. وبالإضافة إلى ذلك أجريت لهؤلاء الأطفال عملية فغر الرغامي كطريقة مؤقتة للتهوية. وتم فطم الأطفال الأربعة بنجاح من تهوية الضغط الإيجابي بعد أن استعيدت وظيفة جانب واحد من الحجاب الحاجز على الأقل.

الكلمات المفتاحية: الشلل الحجابي؛ مرض القلب؛ عمليات الجراحة القلبية؛ طب الأطفال؛ سلسلة حالات؛ عمان.

Bilateral diaphragmatic paralysis (BDP) is a rare condition that can occur after cardiac surgery in children; nevertheless, it constitutes significant morbidity and causes more severe respiratory symptoms than unilateral paralysis.[1,2] Arterial hypoxaemia can result from ventilation-perfusion mismatch at the base of the lungs, while hypercapnia is due to decreased tidal volume and minute ventilation.[3] Cardiovascular surgery is the most common cause of acquired diaphragmatic paralysis, accounting for approximately 64% of phrenic nerve injuries.[4]

The management of BDP involves supporting the respiratory system until the diaphragm recovers enough to ameliorate respiratory distress. As such, BDP almost always requires prompt ventilation using an endotracheal tube, tracheostomy or noninvasive mechanical ventilation (MV).[5] This case series describes the management of four paediatric patients with BDP, emphasising the importance of conservative management until the diaphragm recovers, expected recovery time and the safety of an interventional tracheostomy in such cases.

Case One

A one-year-old male child was admitted to the Royal Hospital, Muscat, Oman, in 2009 with transposition of the great arteries (TGA), a ventricular septal defect (VSD), mild subpulmonic *stenosis* and dextrocardia. He had been born at term via normal vaginal delivery and underwent an uneventful pulmonary artery banding procedure at the age of one month. Following admission, a bidirectional Glenn shunt was inserted with no intraoperative complications. However, during the postoperative period, the child developed low cardiac output syndrome secondary to junctional ectopic tachycardia. He also had malignant hyperthermia and ventilator-associated pneumonia due to an *Acinetobacter* infection.

Initially, the child failed to wean from MV; whenever weaning was attempted, he desaturated and developed tachypnoea and right upper lobe collapse. A bronchoscopy revealed mild right bronchomalacia, which was confirmed by computed tomography angiography. He also began to display withdrawal

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symptoms due to prolonged sedation, although magnetic resonance imaging of the brain was normal. The patient was extubated on the 19th day post-surgery; however, in less than two hours he developed severe respiratory distress and carbon dioxide (CO_2) retention, with a partial pressure of CO_2 ($PaCO_2$) level of 77 mmHg. Following re-intubation, he required minimal MV. Extubation to nasopharyngeal continuous positive airway pressure (CPAP) was re-attempted four days later, after his withdrawal symptoms had been controlled. Although a chest X-ray was normal, this second attempt at extubation also failed. On day 24 following the surgery, fluoroscopy showed BDP, for which he underwent bilateral plication. However, a third extubation attempt to nasopharyngeal CPAP one day later also failed. Another trial of gradual weaning from MV using different modes was also unsuccessful. A tracheostomy tube insertion procedure was therefore performed 43 days after the initial cardiac surgery.

Following insertion of the tracheostomy tube, the patient was gradually weaned from MV; by the 105th postoperative day, he was no longer on MV, there was no evidence of respiratory distress and he was maintaining normal oxygen saturation with an oxygen delivery rate of 1 L/minute. He was decannulated 130 days after the initial cardiac surgery, with no significant complications arising due to the tracheostomy. The patient was transferred from the Paediatric Intensive Care Unit (PICU) to a normal ward on the 133rd day and was discharged from the hospital two days later.

Case Two

A 10-day-old full-term female infant presented to the Royal Hospital in 2010 with TGA and an intact ventricular *septum*. She underwent a successful atrial septostomy on the same day, followed by an uncomplicated arterial switch surgery at 16 days of age. The infant was extubated two days later but soon developed severe respiratory distress, desaturation and respiratory acidosis with a $PaCO_2$ level of 67 mmHg; she was consequently re-intubated. The next day, fluoroscopy revealed BDP.

Unfortunately, 12 days after the cardiac surgery, the patient was accidentally extubated during the provision of standard nursing care and became dusky, desaturated and bradycardic. She was immediately re-intubated. Repeated fluoroscopy and ultrasonography showed persistent BDP on the 15th postoperative day. A tracheostomy tube was therefore inserted two days later and the provision of minimal MV was continued via the tube.

On the 43rd postoperative day, fluoroscopy showed recovery of the right side of the diaphragm only and an attempt at gradual MV weaning failed. Two weeks later, repeated fluoroscopy again showed no evidence of recovery on the left side of the diaphragm. Consequently, the patient underwent plication of the left side of diaphragm 59 days after the cardiac operation. Six days later, MV was discontinued and the patient was successfully decannulated on the 69th postoperative day.

Case Three

A two-day-old full-term female newborn presented to the Royal Hospital in 2011 with TGA and VSD. She underwent a septostomy, with a subsequent arterial switch surgery and VSD closure performed when she was 15 days old. The procedure was complicated by postoperative bleeding requiring re-exploration and haemorrhage control. She was gradually weaned from MV and extubated three days after the surgery, but developed severe respiratory distress within one hour of extubation. She could not maintain oxygen saturation and subsequently developed respiratory acidosis with a pH of 7.1 and $PaCO_2$ level of 81 mmHg. No improvement was noted with noninvasive ventilation and the infant was re-intubated a few hours later. An echocardiogram showed a residual VSD for which the patient underwent surgical closure at 21 days of age. Five days later, a second attempt at extubation failed, with the patient requiring re-intubation within one hour. At 12 and 19 days after the arterial switch operation, fluoroscopy confirmed the diagnosis of BDP. A tracheostomy tube was inserted on the 28 postoperative day.

On the 62nd day following the arterial switch surgery, fluoroscopy indicated good movement on both sides of the diaphragm. However, the patient remained ventilator-dependent due to the following complications. While the diaphragm was recovering, the patient developed pneumonia twice, for which she received full courses of antibiotics. She also developed chylothorax which lasted for 80 days and had poor weight gain. In addition, an episode of infective endocarditis required six weeks of intravenous antibiotics. The patient was eventually transferred from the PICU on the 147th postsurgical day but required a long period of weaning and rehabilitation. Finally, MV was discontinued 210 days after the cardiac surgery and the patient was successfully decannulated four days later.

Case Four

A five-day-old full-term male baby was admitted to the Royal Hospital in 2014 for a type B interrupted aortic arch repair, ligation of the patent *ductus arteriosus* and VSD closure. Postoperative echocardiography indicated that the arch repair was successful, although there was a small residual VSD. He was extubated six days after the surgery but developed respiratory distress and CO_2 retention, with a $PaCO_2$ level of 57mmHg. A chest X-ray showed right lung collapse. After a trial of nasopharyngeal CPAP failed, the patient was re-intubated in under 90 minutes. Fluoroscopy on days eight and 14 confirmed BDP. The patient underwent a tracheostomy on the 18th postoperative day. Three days later, the patient was transferred to a regional hospital in stable condition.

At seven weeks of age, the patient was re-admitted to the PICU at the Royal Hospital for balloon dilatation of an aortic arch re-coarctation. Repeated ultrasound scans showed no movement of the diaphragm on either side. The MV support was continued and the patient was transferred from the PICU. On day 84 post-surgery, ultrasonography and fluoroscopy showed good bilateral diaphragmatic movement. By the 102nd day following the initial surgery, the patient was no longer on MV. However, attempts to decannulate the patient failed. An airway assessment by an ear, nose and throat (ENT) surgeon revealed left-sided vocal cord paralysis and granulomatous tissue above the tracheostomy site. The patient was subsequently discharged on the 163rd postoperative day with a tracheostomy tube *in situ*. Regular follow-up appointments with an ENT surgeon were scheduled. A summary of the demographic and operative features of each case is shown in Table 1.

Discussion

Diaphragmatic paralysis due to phrenic nerve injury is potentially life-threatening in infants.[6] Phrenic nerve injuries due to thoracic surgeries—mainly open-heart surgeries for congenital heart disease—are the most common cause of diaphragmatic paralysis among children.[7] The estimated incidence of unilateral diaphragmatic paralysis following cardiac surgery is 1.31%;[8] however, the incidence of BDP is not clear as most reports are of single cases.[2,9–11] In a study of 867 patients, van Onna *et al*. described 17 cases of postoperative diaphragmatic paralysis (1.9%), of which only one was bilateral (0.1%).[12] A retrospective study by Dagan *et al*. identified nine BDP cases among 3,214 children who had undergone cardiac surgery, resulting in an incidence of 0.28%.[13] Zhang *et al*. reported four cases of BDP out of 47 patients with diaphragmatic paralysis.[14]

In the current case series, a late diagnosis of BDP was made for the first patient. Extubation failed twice before a diagnosis could be made; while the initial extubation failure was attributed to the development of withdrawal symptoms and mild tracheomalacia, the procedure failed again following control of the symptoms and normal chest X-ray findings. A third extubation failure occurred after bilateral diaphragmatic plication. Later, plication was deemed to have been an inappropriate decision due to a lack of experience with such patients. The other three patients in this case series were diagnosed

Table 1: Demographic and operative features of four paediatric patients with bilateral diaphragmatic paralysis following cardiac surgery

Parameter	Case 1	Case 2	Case 3	Case 4
Age	12 months	16 days	15 days	5 days
Gender	Male	Female	Female	Male
Cardiac diagnosis	Congenitally corrected TGA and a large VSD	Simple TGA	TGA and a VSD	IAA and a VSD
Type of cardiac surgery	Glenn shunt	Arterial switch	Arterial switch	IAA repair and VSD closure
Chest left open after cardiac surgery	No	No	No	Yes
Bypass time in minutes	60	160	220	180
Cross-clamp time in minutes	0	76	120	100
Subsequent reoperation	None	None	Residual VSD closure	Balloon dilatation of residual CoA
Intraoperative complications	None	None	Bleeding	None

TGA = transposition of the great arteries; VSD = ventricular septal defect; IAA = interrupted aortic arch; CoA = coarctation of the aorta.

with BDP much earlier, between 3–12 days after the initial cardiac surgery. This timing is similar to that reported by other researchers.[12,13] In cases of BDP, ultrasonography and/or fluoroscopy findings are considered diagnostic.[5,9,10,15–18] All BDP diagnoses in the current series were based on one of these radiological modalities. Imaging was performed following failed extubation attempts or the development of severe respiratory distress soon after extubation and was repeated before the tracheostomy to confirm the diagnosis and to make sure that no early diaphragm recovery had occurred. These imaging approaches were also used subsequently to assess diaphragm recovery and to inform the timing of decannulation and MV discontinuation.

A tracheostomy reduces the risk of an obstructed endotracheal tube, allows immediate oral intake and simplifies the ventilatory weaning process.[15] In the current series, other benefits included reduced sedation and the transfer and management of two patients outside of the PICU. Tracheostomies were performed within 2–6 weeks of the cardiac surgery for all four patients reported in this series. In certain cases, the procedure was delayed so as to allow further extubation attempts and to establish non-invasive nasopharyngeal CPAP. However, subsequent efforts at extubation failed. This indicates that such patients require forms of respiratory support from which they can be weaned upon diaphragm recovery. In the present case series, conservative management allowed the spontaneous recovery of function in at least one side of the diaphragm; hence, this approach is recommended for such patients. If necessary, plication of the contralateral side can be considered, as in the second case. However, plication of both sides of the diaphragm was unsuccessful in the first patient described in this case series. To the best of the authors' knowledge, there are currently no published reports of successful early bilateral plication.

In the four patients described in the current case series, diaphragm recovery took between 6–12 weeks. While this is longer than the maximum recovery time of seven weeks reported by Dagan *et al*. for nine BDP cases, a subgroup of four patients with complicated operative and postoperative courses were reported to require prolonged ventilation for 50–62 days.[13] Three of the patients in the current case series also had complicated postoperative courses, which might explain the delay in diaphragm recovery. In addition, perioperative complications prolonged MV duration, particularly in two patients with multiple postoperative complications. The second patient in the current series had an uneventful postoperative period and the fastest diaphragm recovery time, in which one side of the diaphragm recovered spontaneously and the other side was plicated. She was weaned off MV nine weeks after the cardiac surgery, with removal of the tracheostomy tube a few days later.

In the study by Dagan *et al*., all nine BDP patients underwent MV via a nasotracheal tube until at least one side of the diaphragm had recovered before the contralateral side was plicated; the researchers claimed that using this approach avoided the complications of a short-term tracheostomy.[13] However, intubation and positive pressure ventilation is not without risk, as it increases the likelihood of infection and lung damage and, if heavy sedation is used, augments muscle atrophy and weakness.[19] Kovacikova *et al*. described two patients in which noninvasive positive pressure ventilation was used to manage respiratory failure resulting from BDP; the first patient was fitted with a nasopharyngeal tube and the second with an anesthaesia oronasal mask, with both patients requiring ventilation pressure control.[5] Although the treatment was noninvasive, complications were reported such as pressure sores over the nasal bridge and cheeks and secondary herpetic and chest infections. In addition, the oronasal mask did not fit properly and there was leakage from the nasopharyngeal tube.[5]

In the present case series, accidental decannulation of the tracheostomy tube without loss of airway occurred in one patient; however, there were no other major complications and all of the patients survived. Four episodes of lower respiratory tract infection were observed, with three of them arising in the patient with prolonged chylothorax due to immunoglobulin loss. This patient also had episodes of sepsis, endocarditis and a central line-related infection. All four patients in the current series were discharged without needing further respiratory support. Other published reports of BDP cases have similarly reported no major tracheostomy-related complications.[10,11]

Conclusion

Conservative management allows for the spontaneous recovery of diaphragm function among patients with BDP following paediatric cardiac surgery. However, such patients require ongoing MV support until at least one side of the diaphragm recovers and any respiratory distress is ameliorated. A tracheostomy is recommended as a relatively safe procedure for ventilating BDP patients with no major complications, as observed in the current case series.

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