www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 83.27
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i4.131

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Diaphragmatic Paralysis Following Congenital Cardiac Surgery-A Single Center Experience

Authors
Dr Reyaz Ahmad Lone, Dr Pradeep Bhaskar, Dr Akhlaque Nabi Bhat, Dr Jiju John, Dr Muhammed Riyas K Rahmath, Dr Aslam Faris, Dr Pawel A Tysarowski, Dr Ahmed Elesawy, Dr Shadab Nabi Wani, Dr Faraz Masud, Dr Ahmad Bin Sallehuddin*
Heart Hospital, Hamad Medical Corporation Doha,Qatar
*Section Head Pediatric Cardiac Surgery
Corresponding Author
Dr Reyaz Ahmad Lone
Associate Consultant Pediatric Cardiac Surgery**,** Hamad Medical Corporation, Doha, Qatar
Email: *drreyazlone@gmail.com*

Abstract
Objective - *To study the prevalence and clinical effects of diaphragmatic paralysis following congenital cardiac surgery and to analyse the impact of an aggressive plication strategy in symptomatic patients.*
Type of Study-*Retrospective chart review*
Methods- *27 children who were subjected to either closed or open heart surgery for congenital heart disease and underwent diaphragmatic plication or required prolonged ventilator support for DP post operatively during eight consecutive years from Jan 2006 till Dec 2013 were identified. Medical records of all these patients were retrospectively reviewed for demographics, primary cardiac diagnosis, details of operation, details of diaphragm palsy and management, ventilation days, length of intensive care unit and hospital stay. Any other information which was thought to be relevant was also noted and all data were statistically analysed.*
Results- *The incidence of clinically significant diaphragmatic palsy was 3% with 0.67% frequency of bilateral phrenic nerve palsy. The affected children had a median age of 0.9 months (0.2- 56.9) with a median weight of 3.6kg (2.7-13). 85% of patients were under 1 year with 48% neonates. The median time to diagnosis of DP after primary surgery was 6 days (1-35). Plication was done at a median interval of 7 days (1-38) in unilateral DP patients compared to 30.5 days (21-89) in case of bilateral DP. Following plication the patients were extubated at a median interval of 1day (0-60). The median total ventialtory time (48 v/s 8 days), ICU and hospital length of stay (53.5 v/s 16 and 57 v/s 23 days respectively) were significantly prolonged in case of children with bilateral DP as compared to unilateral DP.*
Conclusion- *Diaphragmatic paralysis following pediatric cardiac surgery continue to be a major cause of morbidity and prolonged hospital stay. Younger children especially infants and single ventricle patients are more affected clinically requiring surgical intervention. An aggressive plication strategy in these subgroup of patients are safe and can promote a faster recovery and shorter hospital stay avoiding complications related to prolonged ventilation.*

Introduction

Diaphragmatic paralysis (DP) following phrenic nerve injury is one of the major complications of congenital cardiac surgery. In current era most common cause of DP is operative trauma which could be attributed to surgical dissection, electro cauterization, stretching, avulsion, use of topical ice slush or cannulation. Redo surgeries pose a higher risk for diaphragmatic palsies due to technical difficulties in dissection caused by fibrous adhesions surrounding the phrenic nerve and the difficulty of identifying the different structures making the nerve more vulnerable to injury. Inability to wean from ventilator, respire-tory distress, atelectasis, recurrent pneumonia and oxygen dependency are the usual clinical presentations of DP. The clinical effect of phrenic nerve palsy can occasionally be very transient and can go undetected especially in older children and adults. It is less tolerated by neonates, infants and young children because of their underdeveloped intercostal musculature, greater chest wall compliance, horizontal rib cage orientation and mediastinal hyper mobility causing a significant impact on their postoperative recovery. Patients with single ventricle physiology depend on a normal respiratory dynamics to maintain effective pulmonary circulation and a reduced respiratory capacity due to diaphragmatic paralysis may not be well tolerated. Traditionally prolonged ventilator support till recovery of the diaphragm had been used to manage DP in symptomatic patients while diaphragm plication is more universally accepted now to facilitate weaning from ventilator support in a timely manner. The timing of surgical intervention is still a matter of debate. The present study was conducted to analyze the outcome of an aggressive approach in symptomatic children with confirmed diaphragmatic palsy after cardiac surgery.

Patient and Methods

All patients who were subjected to either closed or open heart surgery for congenital heart disease and underwent diaphragmatic plication or required prolonged ventilator support for DP post operatively during eight consecutive years from Jan 2006 till Dec 2013 in the study institution were included in the review. The patients were identified from the operating room register by a search for patients undergoing diaphragm plication and a search of intensive care unit register for post-operative cardiac patients with diaphragm palsy requiring ventilator support. A total of 27 patients were identified who met the criteria for inclusion in the study. Medical records of all these 27 patients were then retrospectively reviewed by two physicians. The data collected included age, sex, primary cardiac diagnosis and operation, reason for suspicion and time to diagnose diaphragm palsy, side affected, time to plication, total ventilator days and ventilator days after plication, length of intensive care unit and hospital stay. Any other information which was thought to be relevant was also noted and all data were statistically analyzed.

Management Strategy for treating patients with symptomatic diaphragm palsy

As a routine patients with suspected DP were subjected to ultra sound examination for evaluation of the diaphragm movements and further confirmation was done using fluoroscopy. All patients with unilateral DP with paradoxical breathing who were unable to be weaned off ventilator were treated with diaphragmatic plication. Those with bilateral affection were followed by serial ultrasound 2-3 times a week and whenever there was return of function in one of the hemi diaphragms with paradoxical movement of the other side, the unrecovered side was plicated. Plication was done as soon as the diagnosis was confirmed if possible, and was carried out through thoracotomy via 7^{th} intercostal space on the affected side. Patients who had suspected or documented hemi diaphragm paralyses but remained stable without ventilator assistance, not necessitating surgical intervention were not included in this review.

Statistical Methods

Descriptive statistics in the form of median and range have been calculated for interval variables whereas frequency and percentage were calculated for categorical variables. Data were categorized as unilateral and bilateral diaphragm paralysis groups and compared using non-parametric Mann-Whitney U tests for interval variables and chi-square test for categorical variables. Comparison was also done between neonates and older children within the unilateral DP group. P- value of 0.05 (two tailed) was considered as statistically significant level. SPSS 21.0 package was used for the analysis.

Results

There were 27 cases of diaphragmatic paralysis following cardiac surgery requiring surgical intervention or prolonged ventilator support among 890 major primarycardiac surgical procedures done during the study period.The incidence of clinically significant diaphragmatic palsy was 3% with 0.67% frequency of bilateral phrenic nerve palsy. The median age was 0.9 months (0.2- 56.9). There were 13(48%) neonates, 10 (37%) infants and 4 (15%) children were more than 1 year old. Out of 27 children affected 16 (59.3%) were males and 11 (40.7%) were females. The median weight was 3.6kg (2.7- 13).The primary surgical procedures following which the children developed diaphragmatic palsy is shown in table 1. 4 out of 27 patients developed DP following redo surgical procedures and 5 patients had DP following closed heart procedures (4 coarctation of aorta repairs and one BT shunt through thoracotomy). Left side was affected in 14 patients (52%) and right diaphragm was affected in 7 patients (26%) while in 6 patients (22%) both sides were affected. Median time to diagnosis of the diaphragm was 6 days after primary surgery. The earliest diagnosis of DP was done within 1 day of primary surgery and in one patient the diagnosis was made following readmission with significant eventration of unilateral diaphragm on the follow up visit one month after primary surgery. Children who were symptomatic underwent plication at a median time of 13 days after primary surgery (1-89 days). Following plication the patients were extubated at a median interval of 1 day. 2 patients were not able to be weaned from ventilator due to co-morbidities (one patient was expired on 33rdpost operative day with severe resistant chylothorax and multiorgan dysfunction following arterial switch procedure and a second patient was transferred abroad with necrotizing enterocolitis and cholestatic jaundice following repair of coarctation of aorta and pulmonary artery banding). 56.5 % (13/23) of patients who underwent plication were extubated within 24 hours of plication and 87 % (20/23) patients were extubated within 4 days of placation (2 patients were not extubated and 2 patients didn't require plication). 16 out of the 23 extubated patients required noninvasive respiratory support in the form of high flow nasal cannula, CPAP or BiPAP for a median interval of 1 day (0-17). The median length of ICU stay was 24 days (1-110) and median length of hospital stay was 30 days (5-116). The median total ventilation time including the unilateral and bilateral affection was 11 days (0.1-90).

Unilateral V/s Bilateral diaphragmatic paralysis

There were 6 cases of bilateral diaphragmatic palsy diagnosed during this period with a frequency of 0.67%. All these children were 1 year old or younger with a median age of 3.4 months (0.6-12.3) and a median weight of 5.3kg (3-7). All of them developed bilateral diaphragmatic paralysis following first-time open heart procedures. The median time to diagnose DP was 3.5 days (2-26) as compared to 6 days (1-35) in unilateral cases. 4 of these 6 patients underwent plication upon recovery of unilateral diaphragmatic function at a median time of 30.5 days (21-89) from primary surgery. In case of unilateral DP plication was done at a median time interval of 7 days (1-38). All the plicated patients were

extubated at a median time of 0.8 days (0-33) in case of unilateral DP while the patients with bilateral DP were extubated at a median of 2.3 days (1.8-4) after plication. The median total ventilator time (48 v/s 8 days) , ICU and hospital length of stay (53.5 v/s 16 and 57 v/s 23 days respectively) were significantly prolonged in case of children with bilateral DP as compared to unilateral DP (Table 2). Both the group of patients had similar median noninvasive ventilator day of 1. None of the patients with bilateral DP underwent tracheostomy even though they required prolonged ventilation. But all these children developed ventilator associated pneumonia while on ventilator support.

Table.1 Distribution of diaphragmatic palsy based on the primary surgical procedure.

Primary Surgical Procedure	Number
CoA Repair	4
TOF repair	4
TAPVD repair	4
ASO	4
ASO with CoA repair	1
BT shunt	2
Glenn shunt	2
Nikaidoh procedure	1
Fontan procedure	1
MVR	1
VSD/ AVSD repair	2
ALCAPA correction	1

CoA- *Coarctation of aorta*, ToF- *Tetrology of Fallot*, TAPVD- *Total Anomalous Pulmonary Venous Drainage*, ASO- *Arterial Switch Operation*, BT shunt- *Blalock- Taussig shunt*, MVR- *Mitral Valve Replacement*, VSD- *Ventricular Septal Defect*, AVSD- *Atrio Ventricular Septal Defect*, ALCAPA- *Anomalous origin of Left Coronary Artery from Pulmonary Artery*.

Statistics Based on Median and Distribution

Table.2 Comparison of unilateral and bilateral diaphragm palsies

Variable	**Bilateral DP** Median(*Range*)n=6	**Unilateral DP** Median(*Range*)n=21	**P-value**
Age	3.4(*0.6-12.3*)	0.8(*0.2-56.9*)	0.719
Weight	5.3(*3.0-7.0*)	3.6(*2.7-13.0*)	0.937
Time to diagnose DP	3.5(2.0-26.0)	6.0(*1.0-35.0*)	0.565
Time to Plication	48.0(*21.0-89.0*)	7.0(*1.0-38.0*)	0.072
Ventilation days after plication	2.3(*1.0-4.0*)	0.8(*0.0-13.0*)	0.872
Total ventilation days	48.0(*19.6-90.0*)	7.0(*0.1-44.0*)	0.003
Total NIV days	1.0(*0.0-7.0*)	1.0(*0.0-17.0*)	0.790
Length of ICU stay	53.5(*24.0-110.0*)	16.0(*1.0-65.0*)	0.005
Length of Hospital stay	57.0(*30.0-116.0*)	23.0(*5.0-66.0*)	0.005

Discussion

Paralysis of diaphragm can cause of delayed recovery and excessive morbidity following pediatric cardiac surgery and cardiac surgery is the most common cause of diaphragmatic palsy [(1)]. The incidence of DP diagnosed clinically has been reported to range from 0.28 to 5.4%. The recently reported incidence of diaphragm paralysis by Dagan etal in a series of 3214 patients is 0.28% [(2)], while Joho Arreola et al in their series of 802 patients reported an incidence of 5.4%[(3)]. In a prospective study conducted by Moketal in 50 children undergoing cardiac surgery by direct percutaneous stimulation of phrenic nerve before and after operation showed 10% frequency of phrenic nerve injury which was higher than the previous retrospective reports[(4)]. The incidence of transient or clinically insignificant phrenic nerve palsies detected by electrophysiological measurements may be much higher[(5)]. Certain procedures like arterial switch operation, Fontan procedure and Blalock- Taussig shunt have shown

to have a higher incidence of DP [3]. Our 8 year data showed 3% occurrence of DP which is comparable with other reports in literature.

Neonates and infants are diaphragmatic breathers with intercostal muscles playing little or no role in respiration. Factors like greater chest wall compliance, horizontal rib cage orientation and mediastinal hyper mobility also contribute their vulnerability to respiratory distress and dependency on respiratory support post diaphragm paralysis. In our study nearly half (48%) of the patients were neonates and 85% of the patients who underwent plication were less than 1 year old.Morbidity associated with DP is more significant in smaller infants and neonates. This group of patients require higher index of suspicion and more aggressive management [6]. In patients with fontan physiology, diaphragmatic paralysis causes a reduction or loss of inspiratory augmentation of pulmonary flow. Presence of DP, in these patients leads to increased morbidity, including pleural effusions, ascites, duration of hospital stay, and need for readmissions [7]. This group of patients tends to improve with early diaphragmatic plication[8].

Clinical diagnosis of DP is difficult when the patient is fully supported with ventilator and it is to be suspected when there is unexplained difficulty in weaning patients from ventilator. In spontaneously breathing patients the paradoxical breathing may become more evident. A persistently elevated diaphragm or persistent at electatic changes of the lung base on chest radiograph should raise the suspicion of DP. In our study 13 of 27 patients were diagnosed to have DP following evaluation for failed extubation while 10 patients were unable to be weaned off the ventilator.3 patients were unable to be weaned off non-invasive support following extubation and one patient had persistent lung base atelectasis with oxygen dependency. One infant was readmitted following mild respiratory distress and elevated hemi diaphragm on chest x-ray, one month after primary repair of tetralogy of fallot. Though fluoroscopy has been traditionally considered as the gold standard for diagnosing the DP, recent studies have shown ultrasound to be equally reliable. More over ultrasound offers added advantages of bedside availability, easy and safe repeat examinations and avoidance of radiation exposure [9,10,11]. In all our patients, diagnosis of DP was confirmed by fluoroscopy before plication as part of a departmental protocol. There is still controversy in the management of these patients in terms of requirement of surgical intervention and the timing of intervention. Most of the current reports suggest surgical intervention in smaller patients if symptomatic and remain on ventilator for more than 2 weeks[6,12,13]. Plication of diaphragm has been shown to be safe and useful procedure to improve the ventilation in these patients [14] and video assisted plication has been shown to offer more rapid recovery with less morbidity[15]. However a recent retrospective study concluded that changing to an aggressive strategy with early placation of diaphragm for phrenic nerve injury after pediatric heart surgery was not associated with the better outcome in their experience [16]

Return of diaphragm function after phrenic nerve palsy following cardiac surgery is known to occur but it is difficult to predict the recovery. Smith et al in a recent study has shown that both the plicatedand non plicated patients regain function at similar frequency (60% and 54.8% respecti-vely) following a follow up period from 6 days to 17 years [17]. In a review of 46 patients who had undergone diaphragm plication following cardiac surgery Baker et al have numerically quantitated and demonstrated the return of function with a trend toward improvement over time [18].

Bilateral diaphragmatic paralysis occurs with much less frequency compared to unilateral DP, but affected patients usually do not tolerate ventilator weaning due to severe reduction in lung capacity. In our series there were 6 patients who developed bilateral DP 4 of whom underwent plication of the paralyzed side once there is return of function in one of the hemi diaphragms with paradoxical movement, a management strategy

which was successfully undertaken in a previous report[2]. Plication improved the functional residual capacity of the hemithorax affected by the paradoxical diaphragm movement facilitating timely extubation. Time taken for recovery of function in at least one of the hemi diaphragms varied from 20- 89 days in our patients. Two of these patients with BDP who didn't demonstrate significant paradoxical breathing upon recovery of one of the diaphragms were extubated without plication. Though tracheostomy is occasionally advocated in some patients in order to manage the ventilation whose phrenic nerve recovery is prolonged, it has been reported that tracheostomy following pediatric cardiac surgery can be associated with significant mortality [19]. None of our patients with BDP were tracheostomised even though one of the patients required 89 days for recovery of unilateral diaphragm function. But it was notable that all these patients had developed ventilator associated pneumonia. Use of noninv-asive ventilatory assistance also has been reported as an alternative treatment of bilateral diaphra-gmatic paralysis in infants to avoid tracheostomy or long term endotracheal intubation [20,21]

Limitations

This study is limited by its retrospective nature. There is no control group of patients who were managed without surgical intervention in order to have a true comparison to find out the effectiveness of plication.

Conclusion

Diaphragmatic paralysis following pediatric cardiac surgery continue to be a major cause of morbidity and prolonged hospital stay especially in smaller infants and single ventricle patients. An aggressive plication strategy in these subgroup of patients are safe and effective which can facilitate timely extubation avoiding complications related to prolonged ventilation. Patients with bilateral diaphragm paralysis take variable time to recover with more prolonged ventilation, ICU and hospital stay in comparison to unilateral diaphragm palsy but could be managed effectively by conservative methods.

References

1. Abad P, Lloret J, Martinez Ibanez V, Patino B, Boix-Ochoa J. Diaphragmatic paralysis: Pathology at the reach of the pediatric surgeon. CircPediatr. 2001;14: 21–4.
2. Dagan O, Nimri R, Katz Y, Birk E, Vidne B. Bilateral diaphragm paralysis following cardiac surgery in children: 10 year's experience. Intensive Care Med. 2006;32: 1222–6.
3. Joho-Arreola AL, Bauersfeld U, Stauffer UG, Baenziger O, Bernet V. Incidence and treatment of diaphragmatic paralysis after cardiac surgery in children. Eur J Cardiothorac Surg. 2005;27:53–7.
4. Mok Q, Ross-Russell R, Mulvey D, Green M, Shinebourne EA. Phrenic nerve injury in infants and children undergoing cardiac surgery. Br Heart J. 1991 May;65(5):287-92.
5. Yemisci OU, Cosar SN, Karatas M, Aslamaci S, Tokel K. A prospective study of temporal course of phrenic nerve palsy in children after cardiac surgery. J ClinNeurophysiol. 2011 Apr;28(2):222-6.
6. de Leeuw M, Williams JM, Freedom RM, Williams WG, Shemie SD, McCrindle BW. Impact of diaphragmatic paralysis after cardiothoracic surgery in children.J Thorac Cardiovasc Surg. 1999 Sep;118 (3):510-7
7. Amin Z, McElhinney DB, Strawn JK, Kugler JD, Duncan KF, Reddy VM, et al. Hemidiaphragmatic paralysis increases postoperative morbidity after a modified Fontan operation. J Thorac Cardiovasc Surg. 2001;122:856–62.
8. Ovroutski S, Alexi-Meskishvili V, Stiller B, Ewert P, Abdul-Khaliq H, Lemmer J, et al. Paralysis of the phrenic nerve as a risk factor for suboptimal Fontan hemodyn-

amics. Eur J Cardiothorac Surg. 2005;27:561–5.
9. Miller SG, Brook MM, Tacy TA. Reliability of two-dimensional echocardiography in the assessment of clinically significant abnormal hemidiaphragm motion in pediatric cardiothoracic patients: Comparison with fluoroscopy. PediatrCrit Care Med. 2006;7:441–4.
10. Balaji S, Kunovsky P, Sullivan I. Ultrasound in the diagnosis of diaphragmatic paralysis after operation for congenital heart disease.Br Heart J. 1990 Jul;64(1):20-2.
11. Sanchez de Toledo J, Munoz R, Landsittel D, Shiderly D, Yoshida M, Komarlu R, Wearden P, Morell VO, Chrysostomou C. Diagnosis of Abnormal Diaphragm Motion after Cardiothoracic Surgery: Ultrasound Performed by a Cardiac Intensivist vs. Fluoroscopy. Congenit Heart Dis. 2010 Nov-Dec;5(6):565-72.
12. Mickell JJ, Oh KS, Siewers RD, Galvis AG, Fricker FJ, Mathews RA. Clinical implications of postoperative unilateral phrenic nerve paralysis. J Thorac Cardiovasc Surg. 1978 Sep;76(3):297-304.
13. Hamilton JR, Tocewicz K, Elliott MJ, de Leval M, Stark J. Paralysed diaphragm after cardiac surgery in children: value of plication. Eur J Cardiothorac Surg. 1990;4(9):487-90;
14. Schwartz MZ, Filler RM. Plication of the diaphragm for symptomatic phrenic nerve paralysis. J Pediatr Surg. 1978 Jun;13(3): 259-63.
15. Hines MH.Video-assisted diaphragm plication in children. Ann Thorac Surg. 2003 Jul;76(1):234-6.
16. Georgiev S1, Konstantinov G, Latcheva A, Mitev P, Mitev I, Lazarov S.Phrenic nerve injury after paediatric heart surgery: is aggressive plication of the diaphragm beneficial?Eur J Cardiothorac Surg. 2013 Nov;44(5):808-12.
17. Smith BM1, Ezeokoli NJ, Kipps AK, Azakie A, Meadows JJ. Course, predictors of diaphragm recovery after phrenic nerve injury during pediatric cardiac surgery. Ann Thorac Surg. 2013 Sep;96(3):938-42. doi: 10.1016/j.athoracsur.2013.05.057. Epub 2013 Aug 8.
18. Baker CJ, Boulom V, Reemtsen BL, Rollins RC, Starnes VA, Wells WJ. Hemidiaphragm plication after repair of congenital heart defects in children: quantitative return of diaphragm function over time. J ThoracCardiovasc Surg. 2008 Jan;135(1):56-61.
19. Cotts T, Hirsch J, Thorne M, Gajarski R.Tracheostomy after pediatric cardiac surgery: frequency, indications, and outcomes.J ThoracCardiovasc Surg. 2011 Feb;141(2):413-8.
20. Hoch B1, Zschocke A, Barth H, Leonhardt A.Bilateral diaphragmatic paralysis after cardiac surgery: ventilatory assistance by nasal mask continuous positive airway pressure. Pediatr Cardiol. 2001 Jan-Feb;22(1):77-9.
21. Lubica Kovacikova*, DusanDobos, Martin Zahorec. Non-invasive positive pressure ventilation for bilateral diaphragmparalysis after pediatric cardiac surgery. Interact Cardiovasc Thorac Surg. 2009 Jan;8(1): 171-2.