52 Diaphragmatic Palsy

Vera Bernet

Educational Goals

- To review clinical presentation of diaphragmatic palsy and its pathophysiological mechanism leading to respiratory problems, especially in children of younger age
- To provide a treatment schedule for different age groups

52.1 Clinical Diagnosis and Severity Assessment

Diaphragmatic palsy or paralysis (DP) due to phrenic nerve injury is a rare respiratory condition which may be life-threatening in newborn and young children (Affatato et al. 1988; Haller et al. 1979; de Leeuw et al. 1999). This entity was first described in 1906 in an adult (Stauffer 1979). In newborn patients in the middle of the last century, it was most often due to birth trauma. In the days of advanced neonatal and cardiac surgery, DP is mostly a complication of thoracic surgery (Greene et al. 1975; Schwartz and Filler 1978; Stauffer and Rickham 1972; Stone et al. 1987; Zhao et al. 1985). The incidence is described between 0.3 and 12.8 % and can be unilateral or bilateral (de Leeuw et al. 1999; Schwartz and Filler 1978; Kunovsky et al. 1993; van Onna et al. 1998). DP due to inflammation, neuropathic or idiopathic is rarely seen in this age group.

Clinical manifestation of DP depends on the unilateral or bilateral involvement and the presence of other respiratory problems. In newborn and infants DP most often presents with respiratory distress, presence of thoracoabdominal asynchrony, recurrent atelectasis, pneumonia, inability to wean from the ventilator or reintubation. Older children can compensate the loss of diaphragmatic function and usually present with little or no symptoms when only one side is affected (van Onna et al. 1998; Langer et al. 1988; Mickell et al. 1978; Serraf et al. 1990; Shoemaker et al. 1981; Tönz et al. 1996; Watanabe et al. 1987). Significant respiratory distress in adults is rarely seen if only one side is affected, because accessory muscles of respiration can compensate for the loss of diaphragmatic function. If both sides are affected, the patients present usually with server exertional dyspnoea (Kumar et al. 2004). In children with single-ventricle physiology and Fontan circulation, DP is haemodynamically not well tolerated and may present with cardiac insufficiency, prolonged pleural effusion or ascites (Ovroutski et al. 2005).

With a better rib cage structural stability and ventilatory muscle strength, older children and adults can maximize ventilatory efficiency of a single functioning hemidiaphragm and minimize the paradoxical motion by passively tensing the

V. Bernet
Department of Pediatric Intensive Care and Neonatology, University Children's Hospital Zurich, Steinwiesstr 75, CH-8032 Zurich, Switzerland
e-mail: vera.bernet@kispi.uzh.ch

P.C. Rimensberger (ed.), *Pediatric and Neonatal Mechanical Ventilation*,
DOI 10.1007/978-3-642-01219-8_52, © Springer-Verlag Berlin Heidelberg 2015

paralyzed hemidiaphragm via its attachments to the expanding rib cage (Robotham 1979).

Infants, on the other hand, tolerate diaphragmatic paralysis less well than older children for different reasons. Infants are dependent on diaphragmatic contraction for adequate gas exchange, and if there is unilateral diaphragmatic paralysis, up to 50 % pulmonary function is lost. The intercostal muscles of infants are weaker, and there is a more horizontal orientation of the rib cage, so that DP cannot be compensated by a greater increase in intrathoracic dimension. Another reason is that infants have increased mediastinal mobility. This means that in the presence of DP, the mediastinal contents are shifted to the contralateral side on inspiration, while the paralyzed diaphragm is pulled upward. Pulmonary compliance is decreased on the ipsilateral side, so the diaphragm is unable to resist the negative intrapleural pressure and thus moves paradoxically with each respiration. This results in a significant reduction of the functional residual capacity of the affected and of the normal lung, alveolar collapse and formation of atelectasis. The usual recumbent position of infants and small children may lead to a greater reduction of vital capacity as in older children (Shoemaker et al. 1981; Tönz et al. 1996; Watanabe et al. 1987; Robotham 1979; Mok et al. 1991).

Several imaging modalities have been used to evaluate a diaphragm with suspected DP. The chest radiographs (X-ray) is not useful in its early diagnosis, especially in the neonatal period. Although most often DP is suspected by elevated hemidiaphragm on X-ray or computed tomography, this is not a specific sign because of the wide normal range of the position of the hemidiaphragm (Chetta et al. 2005). Confirmation requires diaphragm mobility tests like fluoroscopy or ultrasound. In the late 1990s ultrasound started to be the diagnostic tool of choice to diagnose DP. Multiple studies could show that ultrasound has advantages over fluoroscopy including portability, lack of ionizing radiation, visualization of structures of the thoracic bases and upper abdomen and the ability to quantify diaphragmatic motion (Alexander 1966; Houston et al. 1995; Gerscovich et al. 2001; Epelman et al. 2005; Miller et al. 2006). In earlier years and still in some hospitals, fluoroscopy was the gold standard to secure diagnosis. Not much intention has been given to the use of magnetic resonance (MR) navigator echo monitoring of the diaphragm so far (Taylor et al. 1999). This might be due to the difficulty to move sick patients and especially children to the MR. Another modality available to confirm the diagnosis of diaphragm paralysis is phrenic nerve stimulation (PNS) and diaphragmatic electromyography (EMG). Both methods are frequently used in adults but difficult to use in infants and newborn. In PNS the nerve is electrically stimulated using percutaneous or needle electrodes or a single posterior cervical coil. EMG is either recorded from cutaneous electrodes between the seventh and ninth intercostal space in the anterior axillary line or from oesophageal electrodes. Mechanical response of the diaphragm is detected and measured by the elicited transdiaphragmatic pressure (Pdi) twitch against a closed airway or compound muscle action potentials. In patients with neuropathy there is usually a prolonged latency and absence or reduced twitch pressure (Luo et al. 2000; Strakowski et al. 2007).

Most important for every examinations is that the patient is under spontaneous breathing. Even ventilation on continuous positive pressure can mask a DP by moving the diaphragm in a lower position. Positioning, quality of movement and paradox movement of the diaphragm should be taken into account.

52.2 Management

Resolution of diaphragmatic paralysis depends on the severity of the phrenic nerve injury. Apart from complete denervation, stretching or blunt trauma of the phrenic nerve usually causes transient phrenic nerve paralysis. In many cases of traumatic injury, normal diaphragmatic function would gradually return over the next 6–12 months, but this would implicate a respiratory support during these times for newborn and small infants.

In children with bilateral paralysis, there is a consensus that one side needs to be plicated.

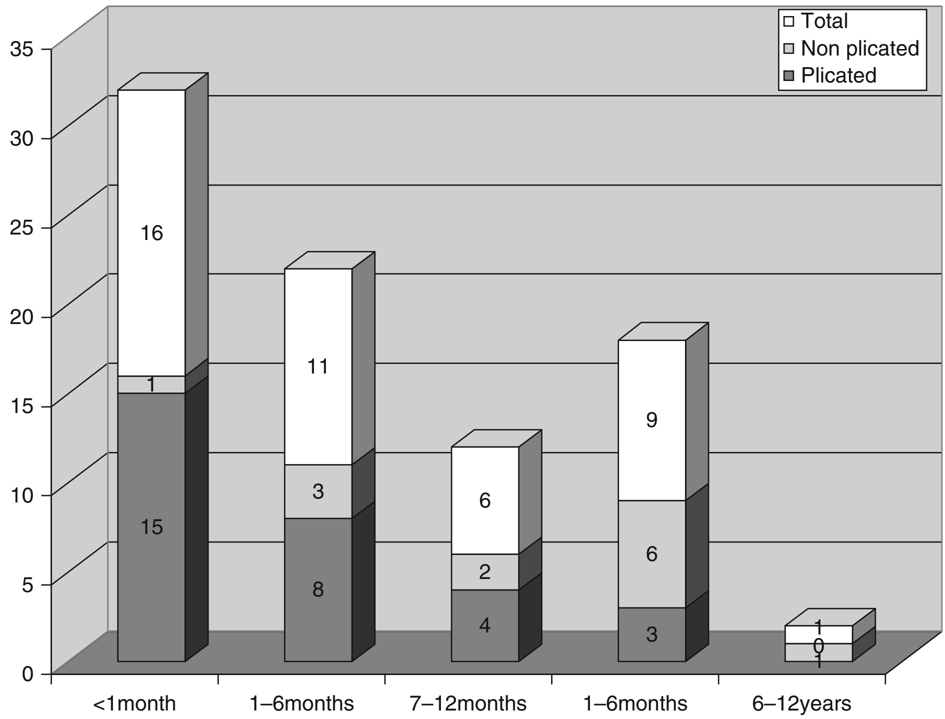

Fig. 52.1 Distribution of age in plicated and non-plicated patients (From Joho-Arreola et al. (2005) with permission)

However, some authors have reported recovery after 2–6 weeks after original operation (Haller et al. 1979; Langer et al. 1988; Mickell et al. 1978; Serraf et al. 1990; Hamilton et al. 1990). Time of intervention and treatment is discussed controversially and depends on the age of the patients.

In a study we published in 2005, we found an incidence of 5.4 % of infants with DP after cardiac surgery. In 42 infants we could show that children older than 6 months tolerated DP quite well, and it is reasonable to wait before plication is performed (Fig. 52.1) (Joho-Arreola et al. 2005). This can happen with noninvasive ventilation (NIV). In infants and older children, only PEEP is needed to reduce work of breathing until the diaphragm recovers (Bernet et al. 2005). This is most often applied either by a nasal or full face mask. Sometimes in small infants and newborns, NIV with patient-triggered positive pressure ventilation is needed to support the patients sufficiently (Robotham 1980).

In young infants under 6 months, plication should be performed after verification of the diagnosis. We developed an algorithm for these patients (Fig. 52.2) (Joho-Arreola et al. 2005). In our institution, we used a thoracic approach as described by Bisgard (Cilley and Coran 1995). In all patients with unilateral paralysis, plication was performed through the seventh intercostal space with a lateral thoracotomy and fixation of the diaphragm on the ventral tenth costal arch. In three cases with bilateral pareses, only one side was plicated similarly with good result. Our results are comparable with the findings of Lemmer et al. which showed in a study of 74 children with congenital heart disease and DP after operation that mechanical ventilation, ICU stay and hospital stay are prolonged in newborns and young infants without plication (Lemmer et al. 2007). Baker et al. performed in 17 patients after DP plication fluoroscopy assessing diaphragm motion. Excursion of the plicated diaphragm was 77 % of the contralateral side. There was a trend toward improved function over time (Baker et al. 2008).

In a follow-up of 22 adult patients, Versteegh et al. could show that the patients after plication

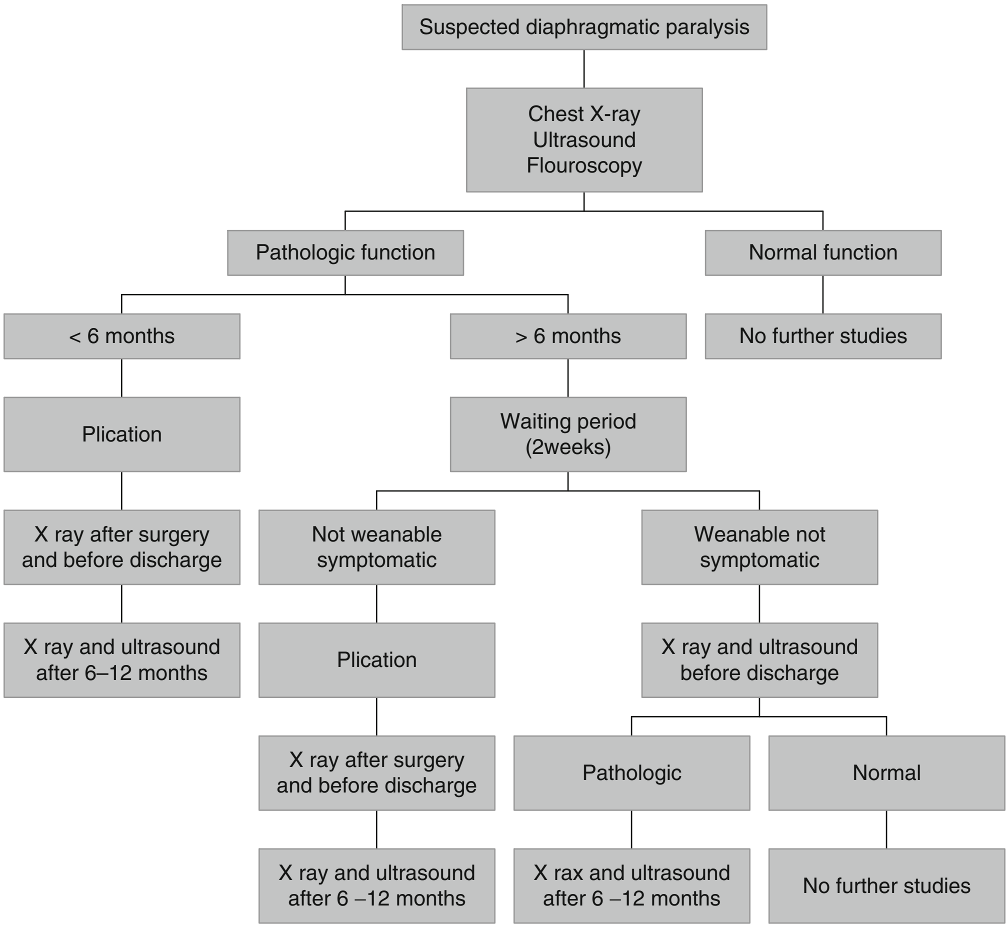

Fig. 52.2 Algorithm of patients with diaphragmatic paralysis (From Joho-Arreola et al. (2005) with permission)

improved in seated vital capacity, supine vital capacity and FEV_1. On the transient dyspnoea index, a remarkable improvement was noted in dyspnoea (Versteegh et al. 2007).

In children undergoing univentricular heart repair with non-pulsatile circulation like after Fontan operation, early diaphragm fixation seems to reduce morbidity like pleural effusions, ascites, duration of hospital stay and need for readmission (Talwar et al. 2010). Video-assisted thoracoscopic plication of the diaphragm has been described in a small case series in children and adults with good results (Freeman et al. 2006; Hines 2003). Hines described five patients between one week and 2 years who were successfully operated with a short ICU stay time (Hines 2003).

Diaphragmatic pacing has been used in adults and older children but not in infants. It requires an intact phrenic nerve and is not an option in bilateral paralysis. It is usually used in children with a nonsurgical origin of paralysis.

Essential to Remember

- Diaphragmatic paralysis is a typical complication after thoracic surgery and should be anticipated if babies cannot get off the ventilator.
- In children older than 6 months, a conservative treatment is eligible.
- Operative treatment is necessary in children under 6 months.

References

Affatato A, Villagra F, De Leon JP, Gomez R, Checa SL, Vellibre D, Sanchez P, Diez Balda JI, Brito JM (1988) Phrenic nerve paralysis following pediatric cardiac surgery. Role of diaphragmatic plication. J Cardiovasc Surg 29:606–609

Alexander C (1966) Diaphragm movements and the diagnosis of diaphragmatic paralysis. Clin Radiol 17: 79–83

Baker CJ, Boulom V, Reemtsen BL, Rollins RC, Starnes VA, Wells WJ (2008) Hemidiaphragm plication after repair of congenital heart defects in children: quantitative return of diaphragm function over time. J Thorac Cardiovasc Surg 135:56–61

Bernet V, Hug MI, Frey B (2005) Predictive factors for the success of noninvasive mask ventilation in infants and children with acute respiratory failure. Pediatr Crit Care Med 6:660–664

Chetta A, Rehman AK, Moxham J, Carr DH, Polkey MI (2005) Chest radiography cannot predict diaphragm function. Respir Med 99:39–44

Cilley RE, Coran AG (1995) Eventration of the diaphragm. In: Smith R (ed) Pediatric surgery. Chapman & Hall Medical, New York, pp 168–175

de Leeuw M, Williams JM, Freedom RM, Williams WG, Shemie SD, McCrindle BW (1999) Impact of diaphragmatic paralysis after cardiothoracic surgery in children. J Thorac Cardiovasc Surg 118(3):510–517

Epelman M, Navarro OM, Daneman A, Miller SF (2005) M-mode sonography of diaphragmatic motion: description of technique and experience in 278 pediatric patients. Pediatr Radiol 35:661–667

Freeman RK, Wozniak TC, Fitzgerald EB (2006) Functional and physiologic results of video-assisted thoracoscopic diaphragm plication in adult patients with unilateral diaphragm paralysis. Ann Thorac Surg 81:1853–1857

Gerscovich EO, Cronan M, McGahan JP, Jein K, Jones CD, McDonald C (2001) Ultrasonographic evaluation of diaphragmatic motion. J Ultrasound Med 20: 597–604

Greene W, L'Heureux P, Hunt CE (1975) Paralysis of the diaphragm. Am J Dis Child 129:1402–1405

Haller JA, Pickard LR, Tepas JJ, Rogers MC, Robotham JL, Shorter N, Shermeta DW (1979) Management of diaphragmatic paralysis in infants with special emphasis on selection of patients for operative plication. J Pediatr Surg 14(6):779–784

Hamilton JRL, Tocewicz K, Elliott MJ, de Leval M, Stark J (1990) Paralysed diaphragm after cardiac surgery in children: value of plication. Eur J Cardiothorac Surg 4:487–491

Hines MH (2003) Video-assisted diaphragm plication in children. Ann Thorac Surg 76:234–236

Houston JG, Fleet M, Cowan MD, McMillan NC (1995) Comparison of ultrasound with fluoroscopy in the assessment of suspected hemidiaphragmatic movement abnormality. Clin Radiol 50:95–98

Joho-Arreola AL, Bauersfekd U, Stauffer UG, Baenziger O, Bernet V (2005) Incidence and treatment of diaphragmatic paralysis after cardiac surgery in children. Eur Cardiothorac Surg 27:53–57

Kumar N, Folger WN, Bolton CF (2004) Dyspnea as the predominant manifestation of bilateral phrenic neuropathy. Mayo Clin Proc 79:1563–1565

Kunovsky P, Gibson GA, Pollock JC, Stejskal L, Houston A, Jamieson MPG (1993) Management of postoperative paralysis of diaphragm in infants and children. Eur J Cardiothorac Surg 7:342–346

Langer JC, Filler RM, Coles J, Edmonds JF (1988) Plication of the diaphragm for infants and young children with phrenic nerve palsy. J Pediatr Surg 23(8):749–751

Lemmer J, Stiller B, Heise G, Hübler M, Alexi-Meskishwili V, Weng Y, Redlin M, Amann V, Ovroutski S, Berger F (2007) Mid-term follow-up in patients with diaphragmatic plication after surgery for congenital heart disease. Intensive Care Med 33:1985–1992

Luo YM, Harris ML, Lyall RA, Watson A, Polkey MI, Moxham J (2000) Assessment of diaphragm paralysis with oesophageal electromyography and unilateral magnetic phrenic nerve stimulation. Eur Respir J 15:596–599

Mickell JJ, Sang O, Siewers RD, Galvis AG, Fricker FJ, Mathews RA (1978) Clinical implications of postoperative unilateral phrenic nerve paralysis. J Thorac Cardiovasc Surg 76(3):297–304

Miller SG, Brook MM, Tacy TA (2006) Reliability of two-dimensional echocardiography in the assessment of clinically significant abnormal hemidiaphragm motion in pediatric cardiothoracic patients: comparison with fluoroscopy. Pediatr Crit Care Med 5:441–444

Mok Q, Ross Russel R, Mulvey D, Green M, Shinebourne EA (1991) Phrenic nerve injury in infants and children undergoing cardiac surgery. Br Heart J 65:287–292

Ovroutski S, Alexi-Meskishvili V, Stiller B, Ewert P, Abdul-Khaliq H, Lemmer J, Lange PE, Hetzer R (2005) Paralysis of the phrenic nerve as a risk factor for suboptimal Fontan hemodynamics. Eur J Cardiothorac Surg 27:561–565

Robotham JL (1979) A physiological approach to hemidiaphragm paralysis. Crit Care Med 7:563–566

Robotham JL (1980) Continuous positive airway pressure in hemidiaphragmatic paralysis. Anesthesiology 52: 167–170

Schwartz MZ, Filler RM (1978) Plication of the diaphragm for symptomatic phrenic nerve paralysis. J Pediatr Surg 13(3):259–263

Serraf A, Planche C, Lacour Gayet F, Bruniaux J, Nottin R, Binet JP (1990) Post cardiac surgery phrenic nerve palsy in pediatric patients. Eur J Cardiothorac Surg 4:421–424

Shoemaker R, Palmer G, Brown JW, King H (1981) Aggressive treatment of acquired phrenic nerve paralysis in infants and small children. Ann Thorac Surg 32(3):250–259

Stauffer UG (1979) Die Relaxatio diaphragmatica im Kindesalter. Angeborene underworbene Formen. Ped Pädol 14:313–327

Stauffer UG, Rickham PP (1972) Acquired eventration of the diaphragm in the newborn. J Pediatr Surg 7(6):635–640
Stone KS, Brown JW, Canal DF, King H (1987) Long-term fate of the diaphragm surgically plicated during infancy and early childhood. Ann Thorac Surg 44:62–65
Strakowski JA, Pease WS, Johnson EW (2007) Phrenic nerve stimulation in the evaluation of ventilator-dependent individuals with C4- and C5-level spinal cord injury. Am J Phys Med Rehabil 86:153–157
Talwar S, Agarwala S, Mittal CM et al (2010) Diaphragmatic palsy after cardiac surgical procedures in patients with congenital heart. Ann Pediatr Cardiol 3:50–57
Taylor AM, Jhooti P, Keegan J, Simonds AK, Pennell DJ (1999) Magnetic resonance navigator echo diaphragm monitoring in patients with suspected diaphragm paralysis. J Magn Reson Imaging 9:69–74
Tönz M, von Segesser LK, Mihaljevic T, Arbenz U, Stauffer UG, Turina MI (1996) Clinical implications of phrenic nerve injury after pediatric cardiac surgery. J Pediatr Surg 31(9):1265–1267
van Onna IEW, Metz R, Jekel L, Wooley SR, van de Wal HJCM (1998) Post cardiac surgery phrenic nerve palsy: value of plication and potential for recovery. Eur J Cardiothorac Surg 14:179–184
Versteegh MI, Braun J, Voigt PG et al (2007) Diaphragm plication in adult patients with diaphragm paralysis leads to long-term improvement of pulmonary function and level of dyspnea. Eur J Cardiothorac Surg 32:449–456
Watanabe T, Trusler GA, Williams WG, Edmonds JF, Coles JG, Hosokawa Y (1987) Phrenic nerve paralysis after pediatric cardiac surgery: retrospective study of 125 cases. J Thorac Cardiovasc Surg 94:383–388
Zhao HX, D'Agostino RS, Pitilick PT, Shumway NE, Miller C (1985) Phrenic nerve injury complicating closed cardiovascular surgical procedures for congenital heart disease. Ann Thorac Surg 39(5):445–449