17 Physical Therapies in Pediatric Respiratory Disease

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Pediatric cardiorespiratory physical therapy (physiotherapy) management spans the spectrum of care from specialist advice to nonpharmacologic interventions for patients with a variety of respiratory conditions. Physiotherapists are an essential part of the multidisciplinary team and physiotherapy is administered from birth to the time of transition to adult services across the continuum of care settings. To provide optimal care, the therapist will clearly identify the indicators for intervention and balance these against the possible risks. Physiotherapy is not a prescribed procedure; the frequency and dosage of therapy is continually adapted and modified in response to identified outcomes and targeted goals.[1,2]

Treating children can be difficult, and the physiotherapist must be responsive to individual needs and have the technical and nontechnical knowledge, skills and attributes to meet these challenges.

General Principles of Physiotherapy

Physiotherapy often focuses on treating or alleviating generic problems that are amenable to intervention rather than being disease-specific (Fig. 17.1). In some instances, however, interventions are selected based upon the underlying disease process (e.g., primary ciliary dyskinesia [PCD] versus cystic fibrosis [CF]), whereby the elements of the mucociliary escalator affected by the disease process may differ (predominantly ciliary dysfunction versus altered sputum rheology). The pediatric respiratory physiotherapist/therapist will perform a wide variety of roles. The professionals and their training vary internationally. In the United Kingdom, physiotherapists are able to treat patients without a referral from a medical doctor and are therefore independent practitioners. Prior to pediatric physiotherapy, informed consent from caregivers and age-appropriate assent from the child are obtained.

The respiratory physiotherapist/therapist needs physiologic knowledge and practical skills to perform a competent respiratory assessment of the child. From this assessment, problems responsive to physiotherapy are identified and treatment strategies are recommended and implemented. Physiotherapists may also assess the reaction to inhaled pharmacologic agents (e.g., bronchodilator response and nebulized antimicrobial bronchoconstriction trials; Chapter 16), provide education in inhaler and nebulizer techniques, and advise on the optimal timing of inhaled medications with respect to sessions of respiratory physiotherapy. They can also assess the need for home oxygen therapy by performing exercise testing with oximetry.[3] Physiotherapists may also help to identify potential causes of respiratory problems (e.g., gastroesophageal reflux [GER] during airway clearance). These issues can then be escalated to be reevaluated by the medical team. If it is felt that pulmonary secretions are a result of aspiration secondary to uncoordinated swallowing, a speech and language assessment is warranted (see section "Aerodigestive Disease").

The timing of physiotherapy treatments can be important; for example, airway clearance should be timed before feeds or delayed for a sufficient time after feeds to avoid vomiting and aspiration. Likewise, physiotherapy should be timed around analgesia when clinically necessary.

Role of Physiotherapy in Pediatric Respiratory Disease

Physical therapies are essential in the removal of excess bronchopulmonary secretions and maintaining and improving exercise capacity. Physical therapy can support ventilation using high-flow nasal cannulas (HFNCs), continuous positive airway pressure (CPAP) or noninvasive ventilation (NIV). Physical therapy should include postural education where appropriate; it can be used to prevent, correct, or improve postural problems, such as kyphosis in CF patients (Fig. 17.2). Postural education can also be helpful in musculoskeletal dysfunction, in children with contractures that inhibit function, or in children with pain that limits range of motion, mobility, and ability to breathe normally. Poor posture leads to tightening of the respiratory muscles, which can lead to chest wall deformity and contribute to a decline in pulmonary function. It is therefore essential that patients with chronic lung disease have a postural assessment and treatment of any musculoskeletal disorders identified, including core muscle imbalance.

Specifically in neuromuscular disease (NMD), physical therapy is essential to maintain ambulation or facilitate standing, where possible, to improve lung function. Optimizing the maturing musculoskeletal and neuromuscular systems of a child with CF may play an important role in the long-term outcome of the child's mental and physical state.[4] It is essential to ask children with chronic lung disease about urinary and fecal incontinence in a private and empathetic setting; reluctance to cough may stem from fear of incontinence. Physical therapies should be directed toward managing this problem.[5,6]

ABSTRACT

Pediatric physical therapy (physiotherapy) is an essential component in the management of many cardio respiratory presentations in infants and children of all ages. The input will vary considerably depending on assessment findings and underlying pathology; however, the general principles remain the same across the spectrum. This chapter covers the key areas of physical therapy for specific conditions such as cystic fibrosis and bronchiectasis regarding airway clearance techniques, exercise, inhalation therapies, ventilator support, and secondary complications. It also identifies the role of cough augmentation and noninvasive ventilation in the management of children with neuromuscular conditions and spinal cord injuries. The role of the physical therapist in the critical care environment is discussed, with strategies modified for the intubated child to assist in secretion clearance and promote early rehabilitation. The importance of postoperative input through mobilization for children that require intervention and adjuncts to support therapy is outlined. Conditions that cause airway or structural abnormalities and may respond effectively to physiotherapy management are discussed. The interventions, as always, are tailored to the individual and monitored for safety and efficacy. Conditions and clinical presentations usually not amenable to physiotherapy are also reviewed. The main focus of the chapter is to highlight the essential elements of pediatric physical therapy for respiratory disease, reviewing the growing evidence base behind the many interventions available in the therapist's "toolbox." Physiotherapy is not only a science but also an art. It requires the therapist to constantly review, modify, and adapt to a potentially changing presentation as well as the ability to communicate with the child and family for engagement and best outcomes.

KEYWORDS

physiotherapy
respiratory therapy
airway clearance of secretions
neuromuscular disease
bronchiectasis
cough exercise

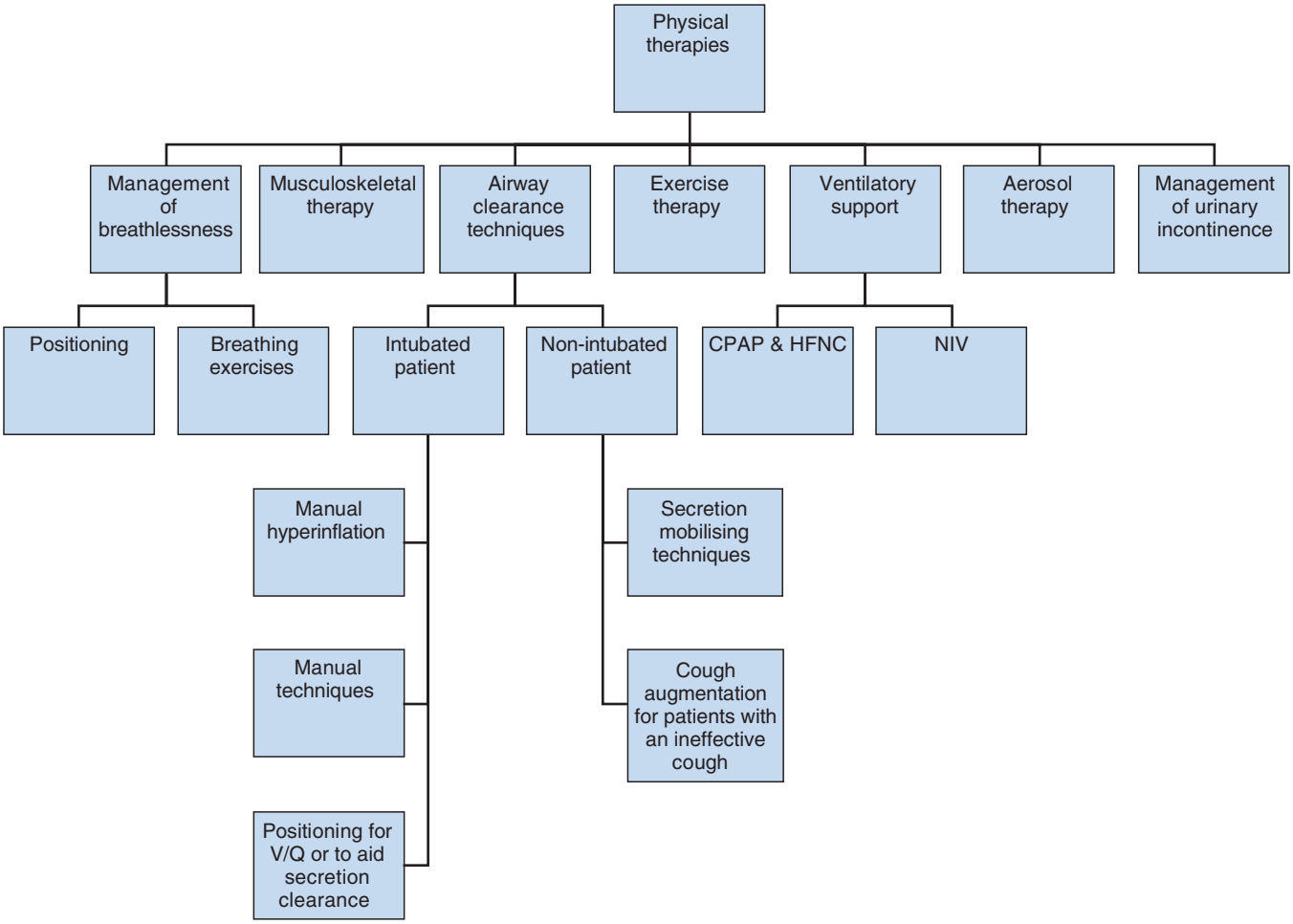

Fig. 17.1 The physical therapies that can be offered to a child with respiratory disease are shown. *CPAP,* Continuous positive airway pressure; *NIV,* noninvasive ventilation; *V/Q,* ventilation perfusion.

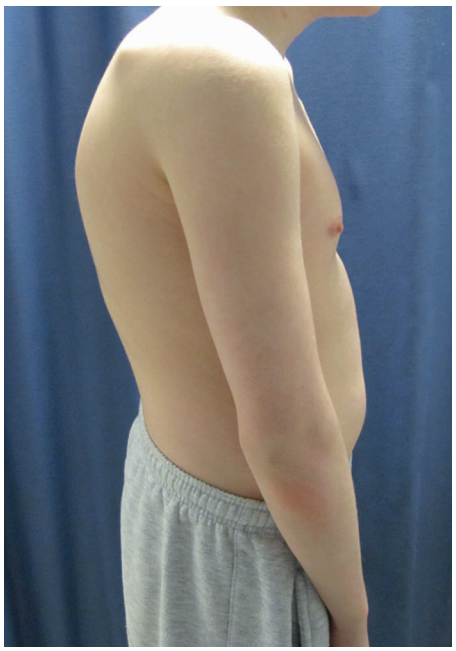

Fig. 17.2 The onset of kyphosis in a male patient with cystic fibrosis.

Respiratory Physiotherapy in Specific Conditions

CYSTIC FIBROSIS AND NONCYSTIC FIBROSIS BRONCHIECTASIS (INCLUDING PRIMARY CILIARY DYSKINESIA)

Airway Clearance

Treatment options for airway clearance depend on the child's age and ability to participate in treatment. There is a wide variety of airway clearance techniques (ACTs; Figs. 17.3–17.5). There is no single best technique, so the therapist should not assume that the findings all apply to non-CF bronchiectasis.[2,7–12]

The technique should be tailored to the individual, and choice is dependent on efficacy, simplicity of use, and cost.[1,2] A good starting point is with the technique that is simplest to use and that impinges least on the patient's life.[13] The term *airway clearance* describes a number of different treatment modalities that aim to enhance the clearance of bronchopulmonary secretions.[14] Through clinical reasoning, the therapist decides the aim of treatment and how to address

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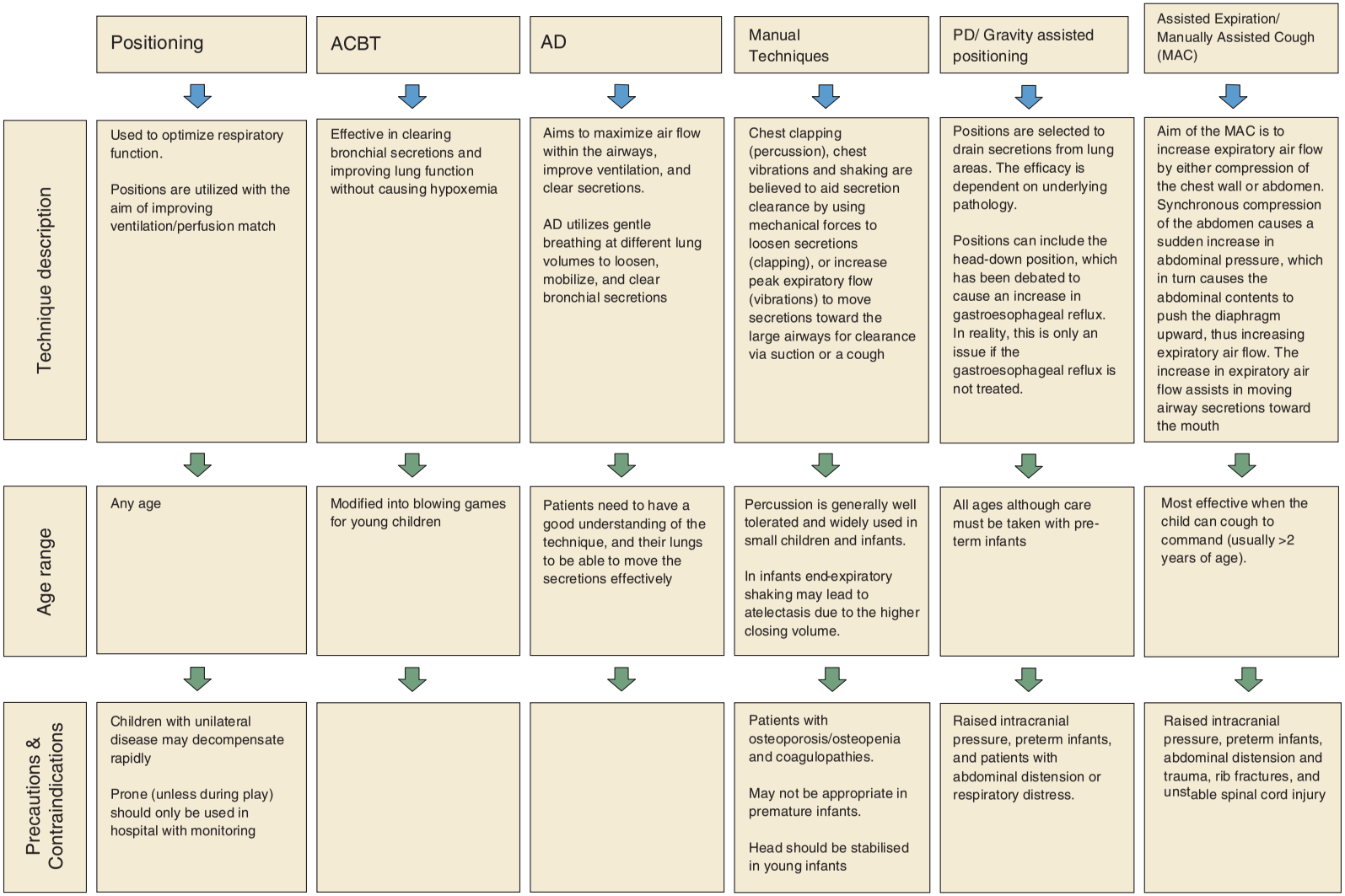

Fig. 17.3 Airway clearance techniques requiring no equipment. Active cycle of breathing techniques: The technique consists of (1) breathing control (BC), which is a resting period of gentle relaxed breathing at the patient's own rate and depth; (2) thoracic expansion exercises, which are 3–5 deep breaths emphasizing inspiration; and (3) forced expiration technique (or "huff"), which combines 1–2 forced expirations followed by a period of BC. The technique is flexible and can be performed in any position. Autogenic drainage (AD): The patient breathes in and holds his or her breath for 2–4 seconds (the hold facilitates equal filling of the lung segments). Expiration is performed keeping the upper airways open (as if sighing). The expiratory force is balanced so that the expiratory flow reaches the highest rate possible without causing airway compression. This cycle is repeated at different lung volumes while collecting secretions from the peripheral airways and moving them toward the mouth. Intermittent positive pressure breathing: Makes include Alpha 200 (Air Liquide Medical Systems, France), NiPPY Clearway (B&D Electromedical, Warwickshire, United Kingdom). Positive expiratory pressure (PEP): Usually PEP consists of a mask with a one-way valve to which expiratory resistance is added. A manometer is inserted into the circuit between the valve and resistance to monitor the pressure, which should be 10–20 cm H_2O during midexpiration. The child usually sits with his or her elbows on a table and breathes through the mask for 6–10 breaths with a slightly active expiration. Makes include PEP Mask (Astratech, Stonehouse, Gloucestershire, United Kingdom), TheraPEP (Smiths Medical, Watford, United Kingdom), Pari PEP (PARI GmbH, Germany). Oscillatory PEP: Positions during use may vary slightly depending on the device type. Often patients will perform 4–8 deep breaths followed by a forced expiration. Makes include the Flutter device (Clement Clarke International Limited, Harlow, Essex, United Kingdom), the Acapella (Henleys Medical, Welwyn Garden City, Hertfordshire, United Kingdom), Aerobika (Trudell Medical Int, Canada). Mechanical insufflation/exsufflation (MI-E): The weaker the child the higher the requirement will be for high insufflation and exsufflation pressures. In children, an insufflation time of less than 1 second is required for equilibration of insufflation pressure and alveolar pressure. Longer exsufflation times do not significantly alter expiratory flows. Higher insufflation and exsufflation pressures both increase expiratory flows, but greater exsufflation pressure had more substantial impact on expiratory flows (1) Cough Assist (Philips Respironics, Andover, Massachusetts) NiPPY Clearway, Pegaso (Dimla-Italia, Bologna, Italy); HFCWO—Makes include Vest (Hill-Rom, St Paul, Minnesota) or Smart Vest (Electromed, New Prague, Minnesota); Intrapulmonary percussive ventilation—Makes include IMPULSATOR—F00012, IPV1C—F00001-C, IPV2C—F00002-C. (Percussionaire Corporation, United States of America), IMP II (Breas, Sweden), Metaneb (Hill-ROM St. Paul, Minnesota). Manual hyperinflation: Patients receive normal tidal volumes coupled with an increased tidal volume using a 500-mL infant bag (or a 1-L bag for older children). A manometer is applied to the circuit to monitor pressures. As a general guide, manual hyperinflation ventilation pressures should not exceed 10 cm H_2O above the ventilator pressure. Flow rates of gas should be adjusted according to the child: 4 L/min for infants, increasing to 8 L/min for children. (From Striegl AM, Redding GJ, Diblasi R, Crotwell D, Salyer J, Carter ER. Use of a lung model to assess mechanical in-exsufflator therapy in infants with tracheostomy. *Pediatr Pulmonol.* 2011;46(3):211-217.)

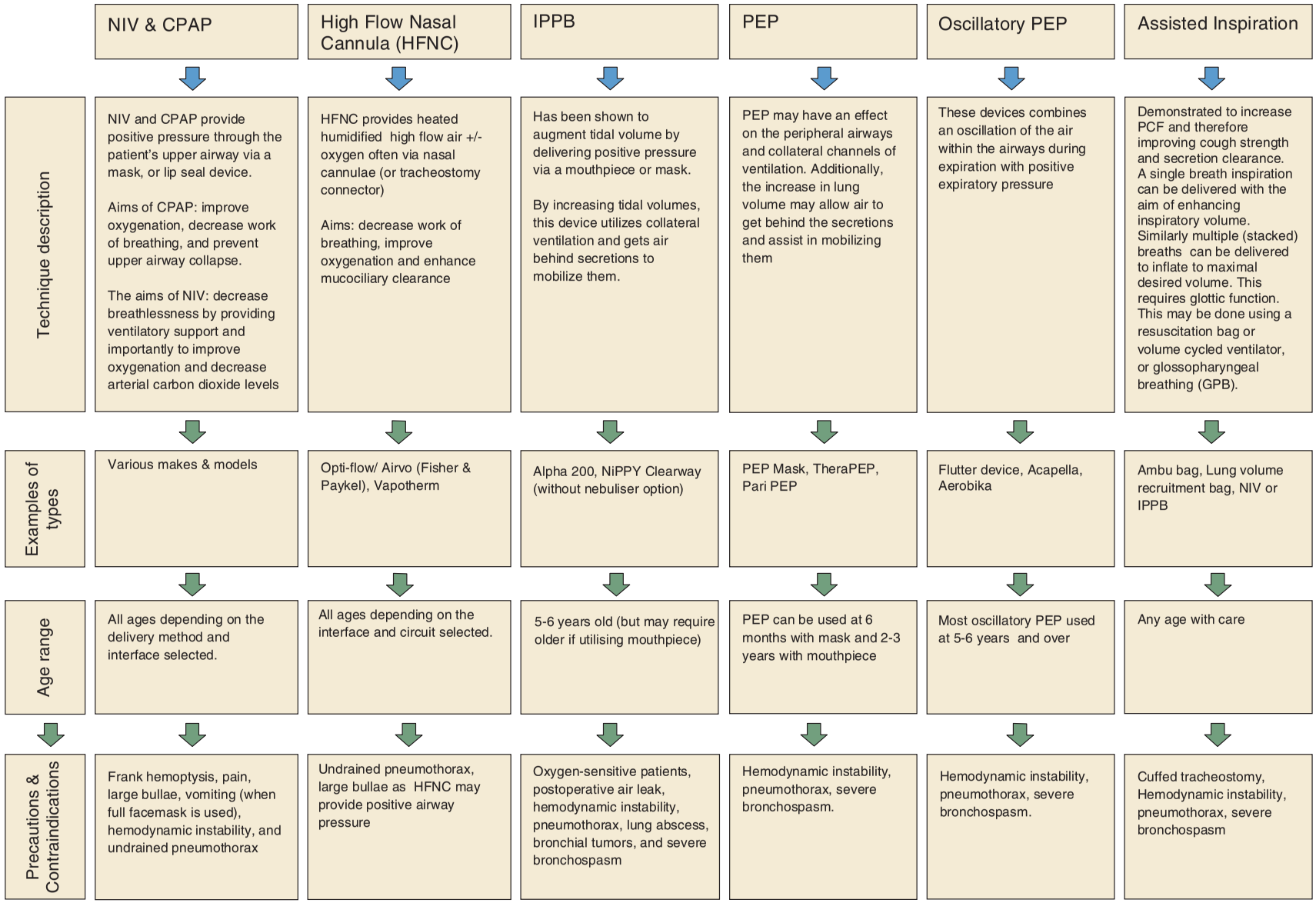

Fig. 17.4 Airway clearance techniques requiring equipment. Airway clearance techniques: The technique consists of (1) breathing control (BC), which is a resting period of gentle relaxed breathing at the patient's own rate and depth; (2) thoracic expansion exercises, which are 3–5 deep breaths emphasizing inspiration; and (3) forced expiration technique (or "huff"), which combines 1–2 forced expirations followed by a period of BC. The technique is flexible and can be performed in any position; Autogenic drainage: The patient breathes in and holds his or her breath for 2–4 seconds (the hold facilitates equal filling of the lung segments). Expiration is performed keeping the upper airways open (as if sighing). The expiratory force is balanced, so that the expiratory flow reaches the highest rate possible without causing airway compression. This cycle is repeated at different lung volumes while collecting secretions from the peripheral airways and moving them toward the mouth. Intermittent positive pressure breathing (IPPB): Makes include Alpha 200 (Air Liquide Medical Systems, France), NiPPY Clearway (B&D Electromedical, Warwickshire, United Kingdom); Positive expiratory pressure (PEP)—Usually PEP consists of a mask with a one-way valve to which expiratory resistance is added. A manometer is inserted into the circuit between the valve and resistance to monitor the pressure, which should be 10–20 cm H_2O during midexpiration. The child usually sits with his or her elbows on a table and breathes through the mask for 6–10 breaths with a slightly active expiration. Makes include PEP Mask (Astratech, Stonehouse, Gloucestershire, United Kingdom), TheraPEP (Smiths Medical, Watford, United Kingdom), Pari PEP (PARI GmbH, Germany); Oscillatory PEP. Positions during use may vary slightly depending on the device type. Often patients will perform 4–8 deep breaths followed by a forced expiration. Makes include the Flutter device (Clement Clarke International Limited, Harlow, Essex, United Kingdom), the Acapella (Henleys Medical, Welwyn Garden City, Hertfordshire, United Kingdom), Aerobika (Trudell Medical Int, Canada).

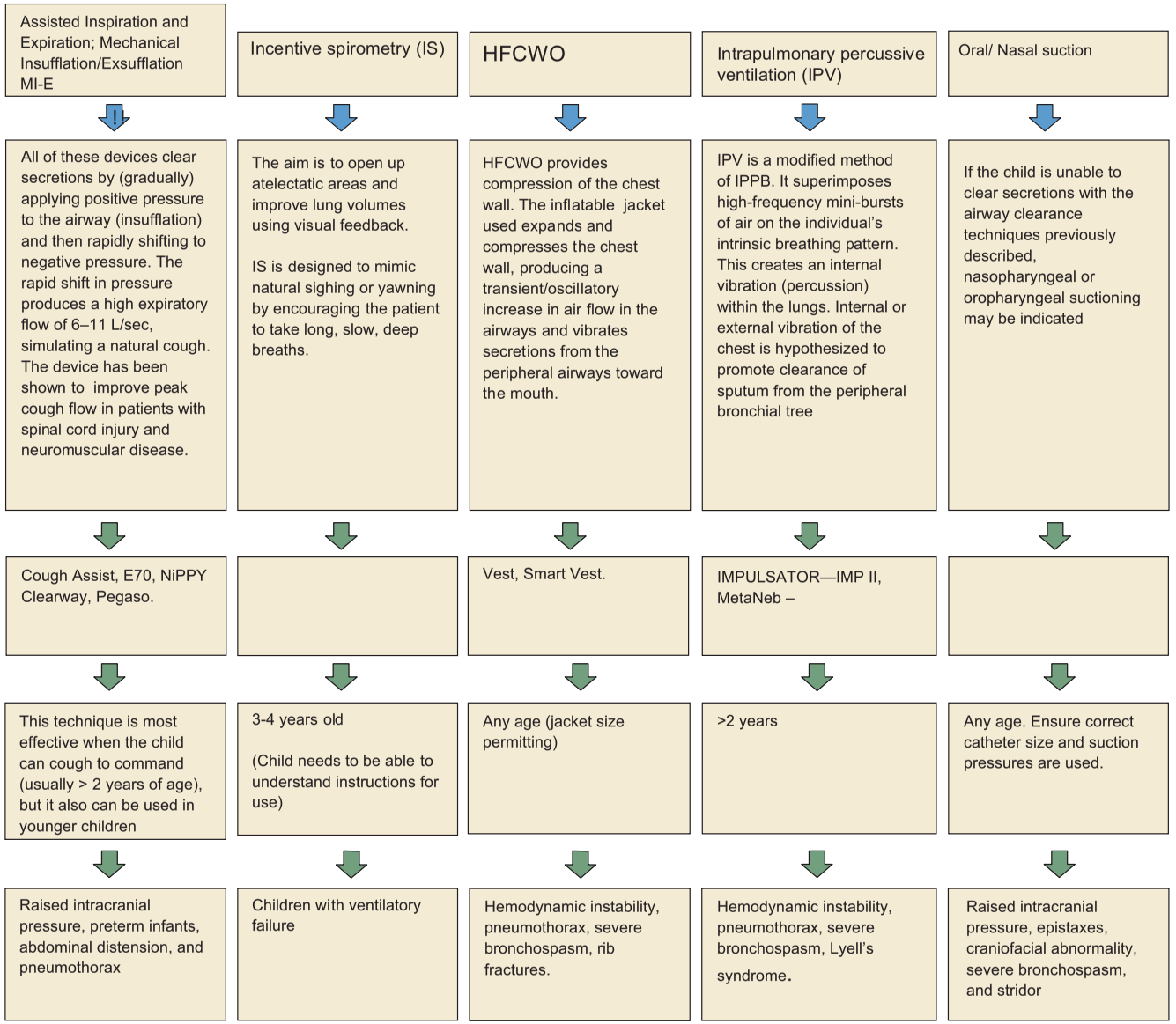

Fig. 17.4, cont'd Mechanical insufflation/exsufflation (MI-E): The weaker the child the higher the requirement will be for high insufflation and exsufflation pressures. In children, an insufflation time of more than 1 second is required for equilibration of insufflation pressure and alveolar pressure. Longer exsufflation times do not significantly alter expiratory flows. Higher insufflation and exsufflation pressures both increase expiratory flows, but greater exsufflation pressure had more substantial impact on expiratory flows (1) Cough Assist (Philips Respironics, Andover, Massachusetts) NiPPY Clearway, Pegaso (Dimla-Italia, Bologna, Italy); High frequency chest wall oscillation—Makes include Vest (Hill-Rom, St Paul, Minnesota) or Smart Vest (Electromed, New Prague, Minnesota); Intrapulmonary percussive ventilation (IPV)—Makes include IMPULSATOR—F00012, IPV1C—F00001-C, IPV2C—F00002-C. (Percussionaire Corporation, United States of America), IMP II (Breas, Sweden), Metaneb (Hill-ROM St. Paul, Minnesota). Manual hyperinflation—Patients receive normal tidal volumes coupled with an increased tidal volume using a 500 mL infant bag (or a 1-L bag for older children). A manometer is applied to the circuit to monitor pressures. As a general guide, manual hyperinflation ventilation pressures should not exceed 10 cm H_2O above the ventilator pressure. Flow rates of gas should be adjusted according to the child: 4 L/min for infants, increasing to 8 L/min for children. *CPAP,* Continuous positive airway pressure; *HFCWO,* high frequency chest wall oscillation; *NIV,* noninvasive ventilation. (From Striegl AM, Redding GJ, Diblasi R, Crotwell D, Salyer J, Carter ER. Use of a lung model to assess mechanical in-exsufflator therapy in infants with tracheostomy. *Pediatr Pulmonol.* 2011;46(3):211-217.)

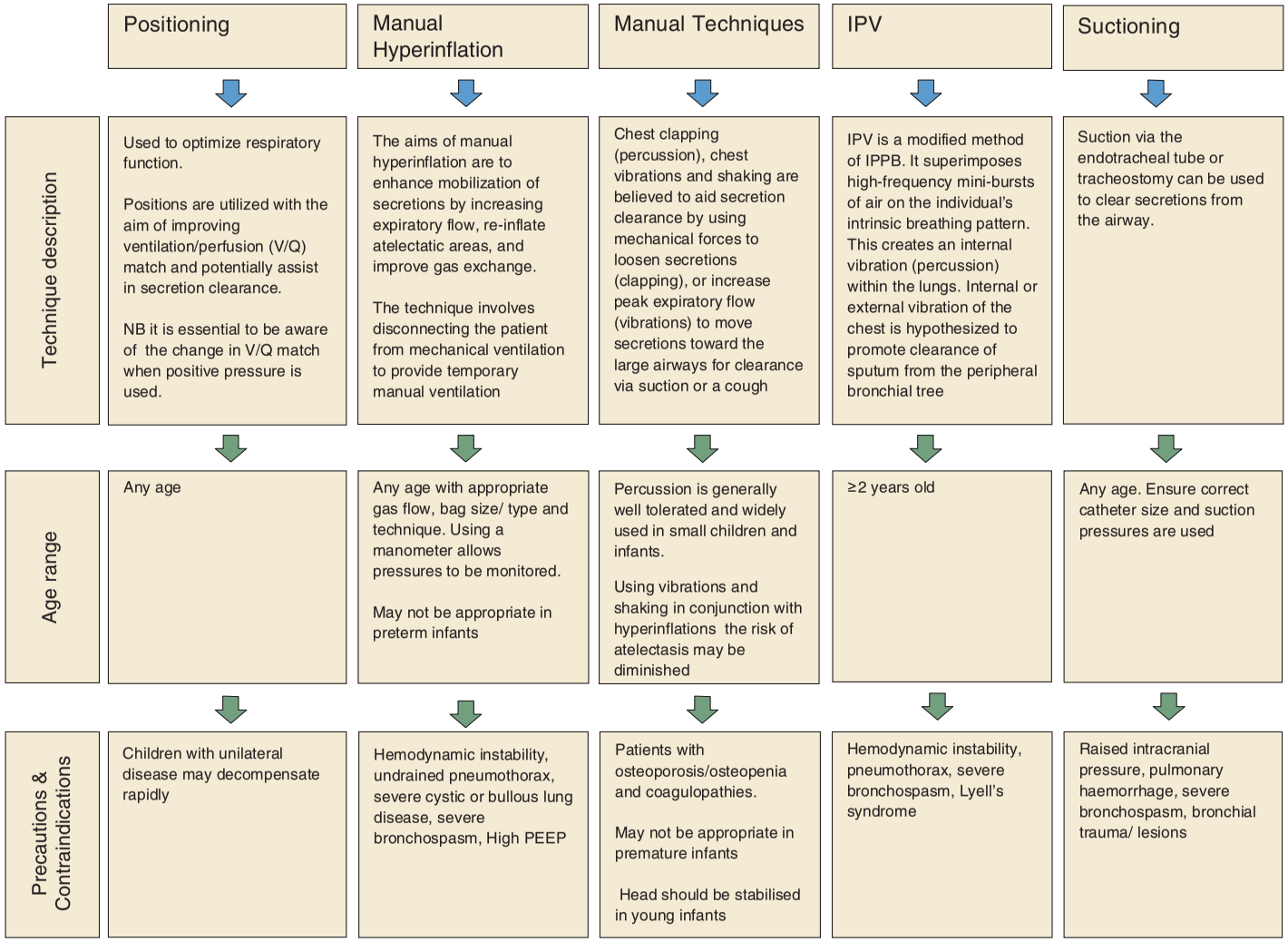

Fig. 17.5 Airway clearance techniques when the patient is intubated. Active cycle of breathing techniques: The technique consists of (1) breathing control (BC), which is a resting period of gentle relaxed breathing at the patient's own rate and depth; (2) thoracic expansion exercises, which are 3–5 deep breaths emphasizing inspiration; and (3) forced expiration technique (or "huff"), which combines 1–2 forced expirations followed by a period of BC. The technique is flexible and can be performed in any position. Autogenic drainage: The patient breathes in and holds his or her breath for 2–4 seconds (the hold facilitates equal filling of the lung segments). Expiration is performed keeping the upper airways open (as if sighing). The expiratory force is balanced so that the expiratory flow reaches the highest rate possible without causing airway compression. This cycle is repeated at different lung volumes while collecting secretions from the peripheral airways and moving them toward the mouth. Intermittent positive pressure breathing: Makes include Alpha 200 (Air Liquide Medical Systems, France), NiPPY Clearway (B&D Electromedical, Warwickshire, United Kingdom). Positive expiratory pressure (PEP): Usually PEP consists of a mask with a one-way valve to which expiratory resistance is added. A manometer is inserted into the circuit between the valve and resistance to monitor the pressure, which should be 10–20 cm H_2O during midexpiration. The child usually sits with his or her elbows on a table and breathes through the mask for 6–10 breaths with a slightly active expiration. Makes include PEP Mask (Astratech, Stonehouse, Gloucestershire, United Kingdom), TheraPEP (Smiths Medical, Watford, United Kingdom), Pari PEP (PARI GmbH, Germany). Oscillatory PEP: Positions during use may vary slightly depending on the device type. Often patients will perform 4–8 deep breaths followed by a forced expiration. Makes include the Flutter device (Clement Clarke International Limited, Harlow, Essex, United Kingdom), the Acapella (Henleys Medical, Welwyn Garden City, Hertfordshire, United Kingdom), Aerobika (Trudell Medical Int, Canada). Mechanical insufflation/exsufflation: The weaker the child the higher the requirement will be for high insufflation and exsufflation pressures. In children, an insufflation time of less than 1 second is required for equilibration of insufflation pressure and alveolar pressure. Longer exsufflation times do not significantly alter expiratory flows. Higher insufflation and exsufflation pressures both increase expiratory flows, but greater exsufflation pressure had more substantial impact on expiratory flows (1) Cough Assist (Philips Respironics, Andover, Massachusetts) NiPPY Clearway, Pegaso (Dimla-Italia, Bologna, Italy); HFCWO—Makes include Vest (Hill-Rom, St Paul, Minnesota) or Smart Vest (Electromed, New Prague, Minnesota); Intrapulmonary percussive ventilation—Makes include IMPULSATOR—F00012, IPV1C—F00001-C, IPV2C—F00002-C. (Percussionaire Corporation, United States of America), IMP II (Breas, Sweden), Metaneb (Hill-ROM St. Paul, Minnesota). Manual hyperinflation: Patients receive normal tidal volumes coupled with an increased tidal volume using a 500-mL infant bag (or a 1-L bag for older children). A manometer is applied to the circuit to monitor pressures. As a general guide, manual hyperinflation ventilation pressures should not exceed 10 cm H_2O above the ventilator pressure. Flow rates of gas should be adjusted according to the child: 4 L/min for infants, increasing to 8 L/min for children. *IPV,* Intrapulmonary percussive ventilation; *V/Q,* ventilation perfusion. (From Striegl AM, Redding GJ, Diblasi R, Crotwell D, Salyer J, Carter ER. Use of a lung model to assess mechanical in-exsufflator therapy in infants with tracheostomy. *Pediatr Pulmonol*. 2011;46(3):211-217.)

the specific issue (or issues). Lannefors and colleagues clearly identified the following four stages of airway clearance; they are the cornerstones to decision making.[15]

1. To get air behind mucus so as to open up the airways
2. To loosen/unstick the secretions from the small airways (Video 17.1)
3. To mobilize the secretions through the smaller airways to the larger airways
4. To clear the secretions from the central airways

The age and adherence of the individual and caregivers as well as disease severity will affect the modalities introduced and in what combination. In the infant, manual techniques[16] (Figs. 17.6–17.8), positioning,[17] infant positive expiratory pressure (PEP[18]; Fig. 17.9), and assisted autogenic drainage (AAD)[19] are used. PEP and AAD focus on enhancing changes in air flow and ensuring the move from "passive" techniques to a more dynamic approach. The use of movement is encouraged from an early age, as it is not only more effective but also more realistic in the younger age group (Video 17.2).

As the child grows older and can become an active participant in therapy, the emphasis will change. The therapist can incorporate techniques that augment volume and introduce the concept of a change in expiratory flow; in many cases, this is a forced expiration.[20] Forced expiration or "huffing" is integral to many techniques and utilizes the theory of the equal pressure point to move mucus to the larger airways.[21] It is also a valuable assessment tool for children, as chest palpation during a "huff" can often be abnormal, with crackles being palpable, even when there are no abnormalities on auscultation. In young children, forced expiration will start as blowing games and then become a more formal component of ACT.

With the child's increasing ability to participate, the active cycle of breathing techniques (ACBT[17,22]; Fig. 17.10) can be taught and may be used in postural drainage (PD) positions (see Figs. 17.3–17.5). Physiotherapy may consist of modified PD targeting the area of lung affected or rotating through different areas to ensure that the lung fields are clear.[16] Other techniques such as autogenic drainage (AD; Fig. 17.11) also can be considered.[23] In addition, many adjuncts are available, with PEP (Figs. 17.12 and 17.13)[24] or oscillatory PEP (Figs. 17.14 and 17.15)[25,26] commonly used to facilitate clearance and to help move the child toward independence if appropriate. PEP has been shown to reduce exacerbations significantly more than other ACTs.[12] However, different

Fig. 17.7 Infant with cystic fibrosis in side-lying position for physiotherapy.

Fig. 17.6 Percussion to the anterior chest for an infant with cystic fibrosis.

Fig. 17.8 Infant with cystic fibrosis in supine position for physiotherapy.

Fig. 17.9 Toddler with cystic fibrosis using mask positive expiratory pressure.

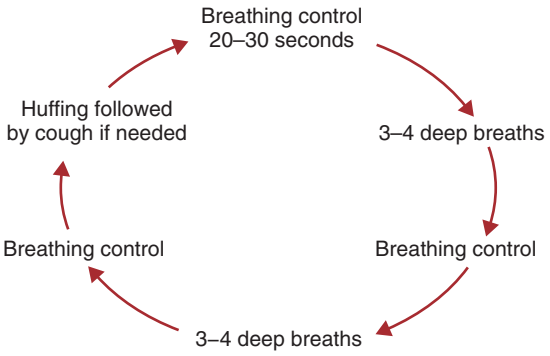

Fig. 17.10 Active cycle of breathing technique.

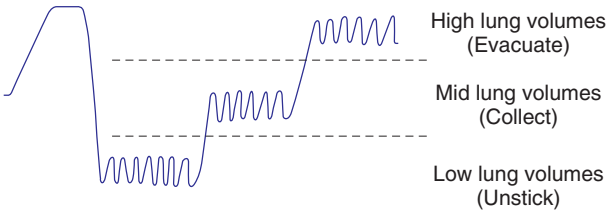

Fig. 17.11 Lung volumes for autogenic drainage.

Fig. 17.12 Girl with cystic fibrosis using PEP (*Pari PEP* [PARI GmbH, Germany]) via a mouthpiece. *PEP,* Positive expiratory pressure.

Fig. 17.13 Mask positive expiratory pressure—Astra PEP (Astratech, Stonehouse, UK).

Fig. 17.14 Oscillatory positive expiratory pressure—Flutter (Clement Clark International, UK).

Fig. 17.15 Oscillatory positive expiratory pressure—Acapella.

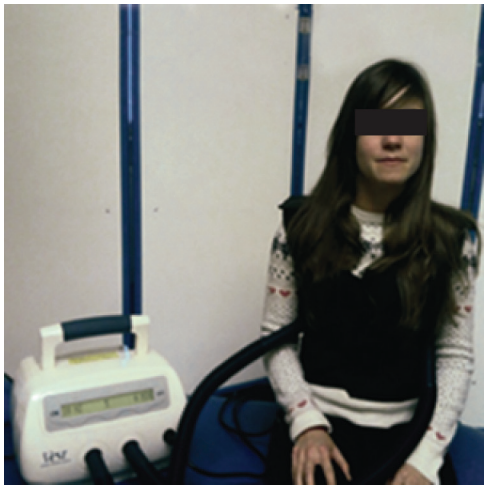

Fig. 17.16 High-frequency chest wall oscillation.

physiotherapy techniques and devices may be more or less effective at varying times (e.g., stable state and during an exacerbation) and in different individuals. CF registry data from 2011 found that oscillating PEP and "huffing" were the most commonly used techniques in the United Kingdom and that PD and high-frequency chest wall oscillation (HFCWO; Fig. 17.16) were the least common.[27] Several of these adjuncts can be used in combination with inhaled medications (e.g., hypertonic saline; Fig. 17.17) or in conjunction with exercise.

HFCWO (see Figs. 17.3–17.5) is widely used across North America. Although evidence indicates that it may be less effective than other therapies,[28,29] including one study showing significantly greater pulmonary exacerbations in patients using HFCWO compared with PEP,[30] it can still be considered for specific individuals or used in combination with other ACTs. One other device that may be of benefit is intrapulmonary percussive ventilation (IPV; Fig. 17.18 and 17.19). Previous studies have investigated sputum mobilization in CF patients by comparing the use of IPV to other modes of airway clearance (e.g., PD and percussion, HFCWO, and oscillatory PEP).[31–33] These studies have shown IPV to be as effective as the other methods of airway clearance in sputum mobilization.

Fig. 17.17 Girl with cystic fibrosis using positive expiratory pressure in combination with nebulized hypertonic saline.

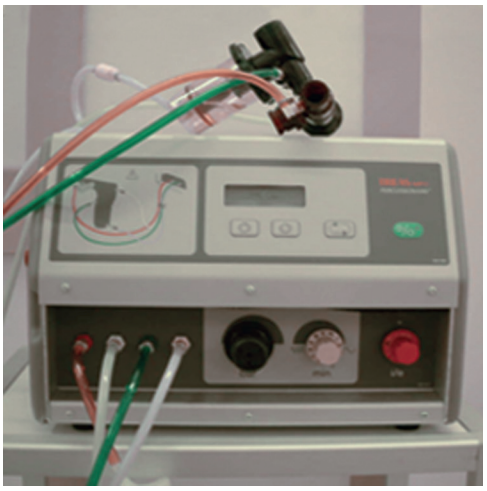

Fig. 17.18 Intrapulmonary percussive ventilation.

Fig. 17.19 The MetaNeb® System (Hill-ROM, St. Paul, Minnesota).

Fig. 17.20 Teenager with cystic fibrosis exercising on a "trampet."

Exercise has many benefits for people with CF and bronchiectasis: improving cardiovascular fitness, bone mineral density, and quality of life (Fig. 17.20). In CF it has been shown to slow the rate of pulmonary function decline[34] and may increase survival independent of FEV_1.[35] Exercise has also been shown to have an additive effect on sputum production, and it improves oxygen saturation in adolescents and adults with CF when used before airway clearance.[36] In fact, Dwyer and coworkers[37] have shown that exercising on a treadmill increases expiratory air flow and moves sputum from peripheral lung regions, but this must be combined with "huffing" to be an effective method of airway clearance.

Regular review of ACT is advised to ensure continuing effectiveness and adherence with therapy; appropriate adjustments to treatment can be made as necessary.[38,39] It is vital to monitor adherence, as, for example, poor adherence in bronchiectasis has been shown to affect important health outcomes including pulmonary exacerbations.[40] The most effective technique may not be the optimal strategy for the individual; multiple contributing factors must be considered. The regimen must be specific to each individual's changing needs and preferences, and as patients grow older, their understanding of the need for the treatment and of its goals must be clear. ACTs have been shown to have short-term effects on mucus transport, but the long-term effects are less clear.[41] There is debate worldwide regarding the introduction of ACT prior to diagnosis of bronchiectasis. There is consensus on the symptomatic patient, where response to treatment is evident; however, there is less agreement on the asymptomatic patient.[14] Chest physiotherapy does not have to be routinely performed unless the underlying diagnosis affects the normal mechanisms of airway clearance. In CF, there is evidence that inflammation, infection, abnormal lung function and ventilation inhomogeneity are present early in life.[42] In addition, for CF or PCD, it seems unethical to wait for airway damage to occur, as the child will thus never have normal mucociliary clearance.[43]

Fig. 17.21 Girl with cystic fibrosis using ultrasonic nebulizer for saline inhalation.

Ventilatory Support

As the disease progresses, use of devices that provide some inspiratory ventilatory support may be the treatment of choice. The use of NIV has been shown to significantly improve ease of sputum clearance, reduce work of breathing,[44] facilitate inspiration and correct respiratory failure; it is widely used in the adult setting. It has also been shown to significantly increase FEV_1 and reduce fatigue in CF adults hospitalized with an acute exacerbation.[45] However, a review of its use in CF children[46] demonstrated a need for more evidence and protocols to identify indications for this age group.

Inhalation Therapies

Effective treatment in this group of patients might have to be supported by inhaled therapies (Fig. 17.21) such as bronchodilators; mucoactive agents such as hypertonic saline, dry powder mannitol (Bronchitol, licensed for over 6 years in Australia and 18 years in Europe) and deoxyribonuclease (RhDNase); antimicrobials, and, where indicated, oxygen therapy.[47–49] The timing of inhalation therapy around airway clearance is important (Fig. 17.22). For example, bronchodilators and mucokinetics should be given before or during ACT to prepare the airways.[50] RhDNase can be administered before (minimum 30 minutes) or after ACT to suit the individual, although in children with well-preserved lung function FEF_{25} was improved if RhDNase was given prior to ACT.[51] If RhDNase is given before ACT, it should be a longer time interval than immediately before ACT; in children it may be given before bed.[52] Antimicrobials and inhaled steroids should be administered following ACT.

Complications and Implications for Physical Therapy

The relationship between ACT (particularly gravity-assisted positions) and GER remains unclear.[53,54] Physiotherapists must be aware of the possible risks with treatment and modify their therapies accordingly; thus the decision should not be

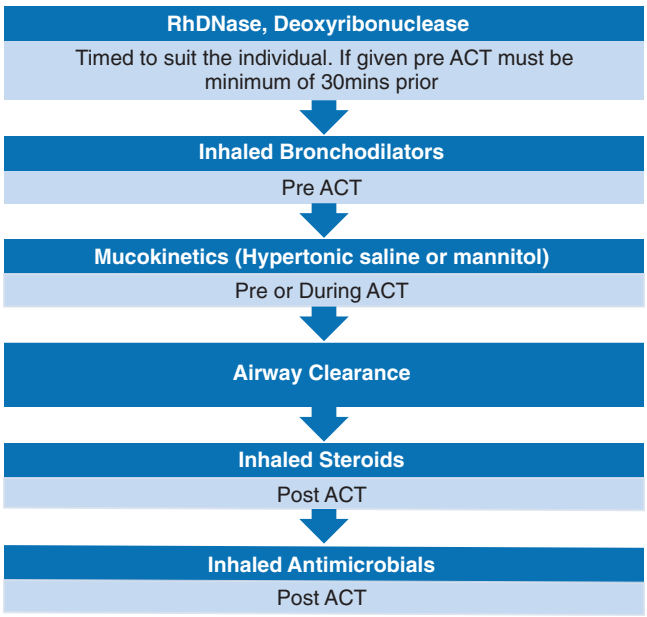

Fig. 17.22 The timing of inhaled therapies around physiotherapy. The algorithm is individualized to the patient and will not necessarily take all of the inhaled therapies listed. *ACT,* Airway clearance technique.

on whether gravity will induce reflux but whether it is necessary to tip at all. Few therapists would tip a 4-week-old asymptomatic CF infant, but in a 1-year-old child with PCD that is at risk of developing acute large airway mucus impaction and lobar changes, PD may well form part of the treatment.

As mentioned, the impact of musculoskeletal issues including posture, incontinence,[55] and pain should be assessed and managed appropriately, including limiting muscle imbalance and strength training to improve the mechanics of breathing and overall well-being.[56] In addition, with the increasing longevity of patients and comorbidities such as diabetes, osteopenia will have to be considered by the therapist and management modified responsively. Often the combination of treatments can be beneficial; for example, ACT using PEP while the patient is on a therapy ball, utilizing core muscle strengthening can provide enhancements as well as reduce treatment time.

RESPIRATORY MUSCLE WEAKNESS

Neuromuscular Disease

The ability to clear bronchopulmonary secretions is essential to prevent sputum retention and associated complications, including lower respiratory tract infection. An effective cough is a vital mechanism to protect against respiratory tract infections, which are the commonest causes of hospital admission in patients with respiratory muscle weakness from NMD.[57] An intact afferent and efferent pathway is required for an effective cough. Reduced airway sensation will fail to elicit a cough in response to a noxious stimulus. Diseases of the nerves (upper or lower motor neuron), neuromuscular junction, or muscles may impair the efferent pathway.

The act of coughing involves the following three main components[58]:

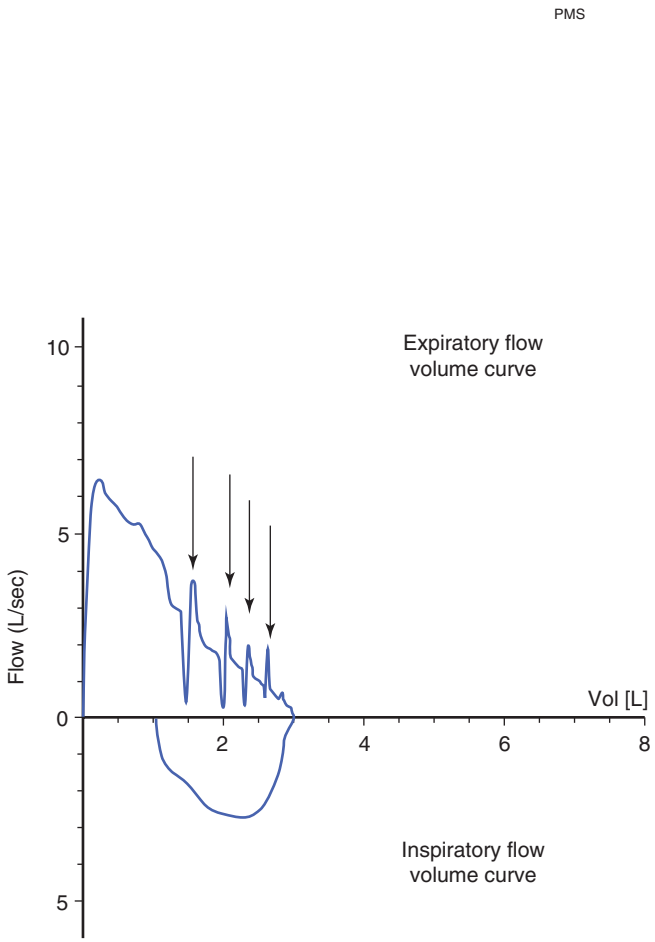

Fig. 17.23 A series of cough spikes is superimposed on the maximal expiratory flow. Cough flow spikes are highlighted *(arrows)*.

1. Deep inspiration up to 85%–90% of total lung capacity
2. Glottic closure, which requires intact bulbar function so a rapid closure of the glottis occurs for approximately 0.2 seconds
3. Effective contraction of the expiratory muscles (abdominal and intercostal) to generate intrapleural pressures of more than 190 cm H_2O.

If one or more of these three components is impaired, the cough will be less effective[59] and the individual may be unable to produce the transient flow spikes essential for an effective cough (Fig. 17.23).[60] Cough strength can be measured by peak cough flow (PCF). PCF is the result of an explosive decompression that generates a flow rate as high as 360–944 L/min in children older than 12 years of age.[61] It is important not to quote adult PCF threshold values for children younger than 12 years of age and to use the appropriate quartile reference value.[61] Physical therapies involve the assessment of cough efficacy by formally evaluating the inspiratory, glottic and expiratory components of the cough along with audibility and measuring PCF with a peak flow meter attached to a face mask.

Poponick and coworkers[62] demonstrated that acute viral illness was associated with a reduction in vital capacity (VC) due to reduced inspiratory and expiratory respiratory muscle strength (by 10%–15% of baseline values). This reduction in

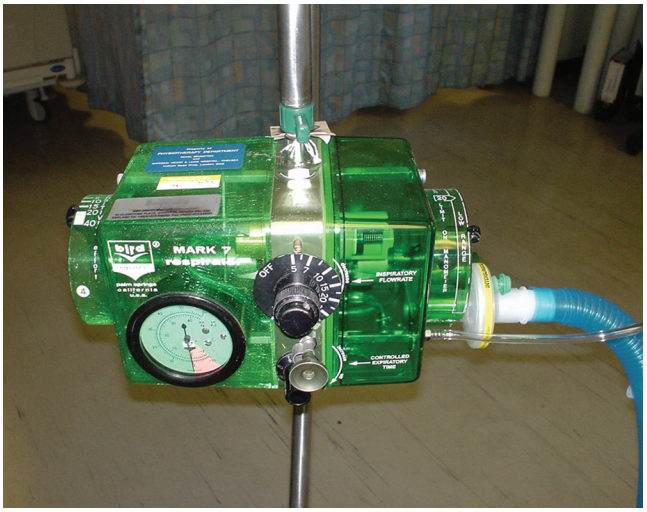

Fig. 17.24 Intermittent positive pressure breathing device.

Fig. 17.25 Lung volume recruitment bag (Intersurgical, Wokingham, England, UK) for the use in assisted inspirations.

inspiratory and expiratory muscle strength will also cause a decline in PCF, possibly to the critical level where the child is unable to clear secretions alone or with his or her normal assisted cough technique. Assisted cough techniques should be targeted to whichever component of the cough is reduced and may consist of assisted inspiration: single breaths (inspiration with expiration between) given with an Ambu bag or an intermittent positive pressure breathing (IPPB) device (Fig. 17.24).[63] They may also consist of stacked breaths (no breath out between inspirations) performed with an Ambu bag (Fig. 17.25), a lung volume recruitment bag, volume-cycled NIV,[64] or glossopharyngeal breathing (GPB).[65] Assisted cough techniques may also be applied during expiration, as with a manually assisted cough (Fig. 17.26; see also Figs. 17.3–17.5). If there is both inspiratory and expiratory muscle weakness, then a combination of assisted inspiration and expiration techniques should be utilized. These techniques have been shown to increase PCF either alone or in combination.[66–69]

Fig. 17.26 Manually assisted cough for a patient with neuromuscular disease.

If the cough is extremely impaired, the patient may require mechanical insufflation/exsufflation (MI-E) (Fig. 17.27).[70,71] This procedure clears secretions by gradually applying a positive pressure to the airway (insufflation) and then rapidly shifting to negative pressure (exsufflation). The rapid shift in pressure produces a high expiratory flow, simulating a natural cough. In pediatric patients with NMD, MI-E has been provided as part of a protocol in patients with a PE_{max} of 20 cm H_2O or less.[72] MI-E has also been shown to expedite secretion removal[73] and prevent intubation.[74] A combination of NIV and MI-E in spinal muscular atrophy type I has been associated with increased life expectancy when compared with the untreated natural history in observational studies.[75–78]

NIV (Fig. 17.28) may also have a role in preventing chest wall deformity in NMD.[78,79] Physical therapies that consist of secretion-mobilizing techniques in NMD may need to provide ventilator support. Such options include IPV (see Figs. 17.3–17.5),[80–82] a modified ACBT (adjusting the patients NIV settings), or HFCWO[83] on NIV where indicated (see Figs. 17.3–17.5). Patients who use NIV should not be taken off their ventilator for physiotherapy or transferred to CPAP. Some patients with NMD are provided with NIV to decrease their work of breathing and augment tidal volumes to enhance ACT. The ACBT can be modified by using NIV to provide deep breaths that mobilize secretions with or without manual techniques, followed by an assisted cough technique.

Outcome is improved when children with NMD are treated with an oximetry-driven protocol when they are unwell to avoid intubation or in preparation for extubation.[84] The protocol consists of carrying out cough augmentation techniques when the oxygen saturations decline below 95% on room air or on NIV without supplementary oxygen therapy. The rationale is that patients have atelectasis with secretion retention and treatment is required to reverse the process; this has been shown to increase survival in patients with NMD. Performing cough augmentation when the patient's oxygen saturation has declined below 95% has contributed to successful extubation and prevented the need for intubation in this patient group.[57,75,78,85]

Currently there is no evidence base for the use of respiratory muscle training in this patient group.[84]

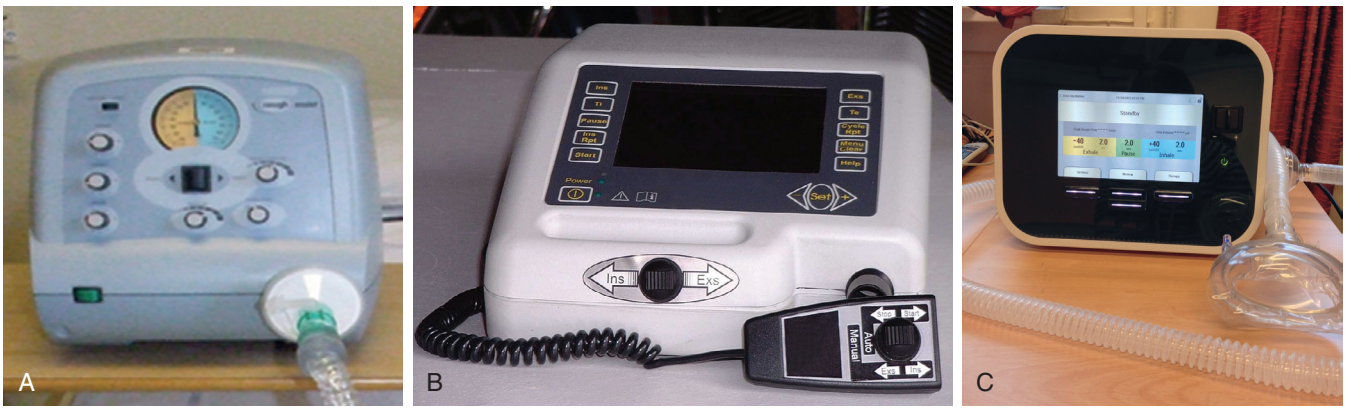

Fig. 17.27 Cough assist device.

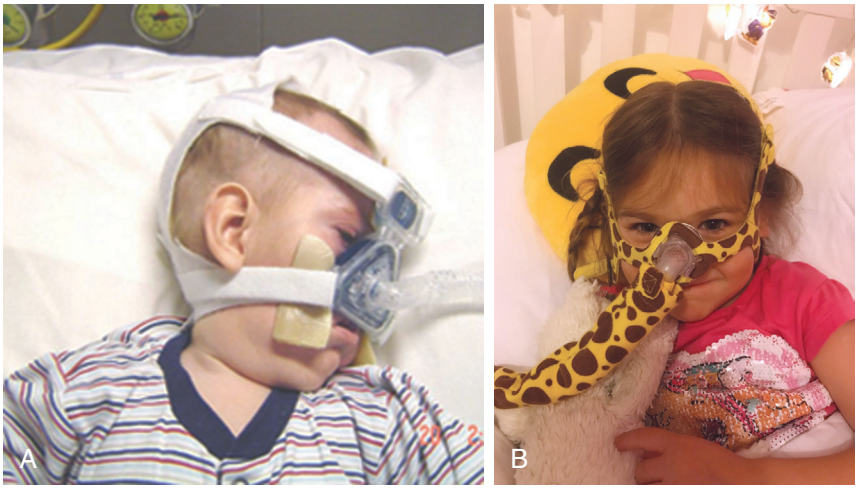

Fig. 17.28 Child with noninvasive ventilation via nasal mask.

Spinal Cord Injury

Physical therapies should be appropriate to the level of the spinal cord injury. Cervical spinal cord injuries may require ventilator support in the form of NIV or, in higher spinal cord injuries, tracheostomy with intermittent positive pressure ventilation (TIPPV). Patients may need to be taught lung volume recruitment maneuvers in the form of assisted inspirations (see section "Neuromuscular Disease" and Figs. 17.3–17.5) to promote chest wall stretching,[86] along with lung growth and development. For injuries that included the thoracic spine, MAC should be taught (see Chapter 72 and Figs. 17.3–17.5).[87] Patients who are significantly weak are likely to benefit from MI-E either via a mask or tracheostomy; this has been reported to be less uncomfortable than suctioning.[88] Abdominal binders may also be required to minimize the effect of postural hypotension and aid in respiration.[89,90]

SURGICAL AND CRITICAL CARE

Preoperative and Postoperative Management

Routine preoperative therapy is not indicated[91]; however, it may be desirable to teach physical therapies to high-risk children prior to surgery—for example, patients with coexisting respiratory conditions that may predispose to postoperative respiratory atelectasis and infection or patients with reduced mobility.

Exercise therapy is an essential part of postoperative care, and endurance training has a favorable influence on pulmonary function in patients after surgical correction of scoliosis.[92] Other postoperative physical therapies differ depending on the patient's status. Breathing exercises such as ACBT may be appropriate (or blowing games in younger children) if secretion retention is an issue. Incentive spirometry should not be used routinely[93] or if the child is mobilizing sufficiently; however, in isolated cases it can give visual feedback and encourage the child to take deeper breaths in an attempt to resolve atelectasis.[94] If there is a significant amount of atelectasis with a decline in oxygen saturation, a step up to HFNC,[95] CPAP or NIV may be necessary. IPPB (Bird Mark 7, Viasys Health Care, Bilthoven, The Netherlands)[96] may be of benefit for older children with sputum retention (see Fig. 17.24).[63,97]

Critical Care

The physiotherapist is an integral part of the intensive care team, the indicators for intervention must be clearly defined and possible adverse events to these recognized. In the

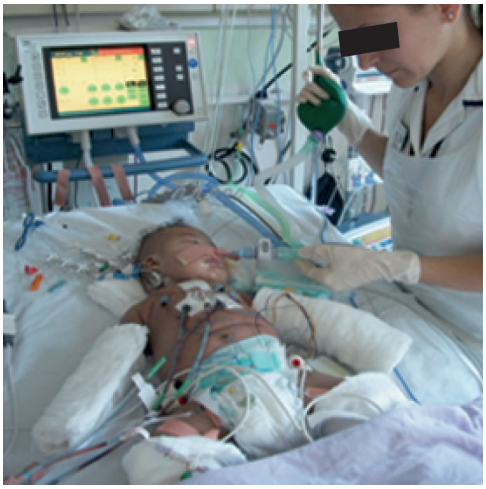

Fig. 17.29 Physiotherapy (manual hyperinflation and manual techniques) being performed on a intubated and ventilated infant.

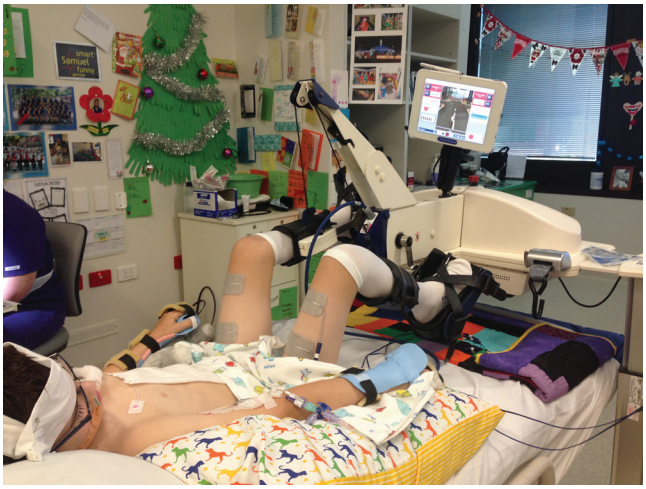

Fig. 17.31 Functional electrical stimulation being used to assist rehabilitation for the lower limbs.

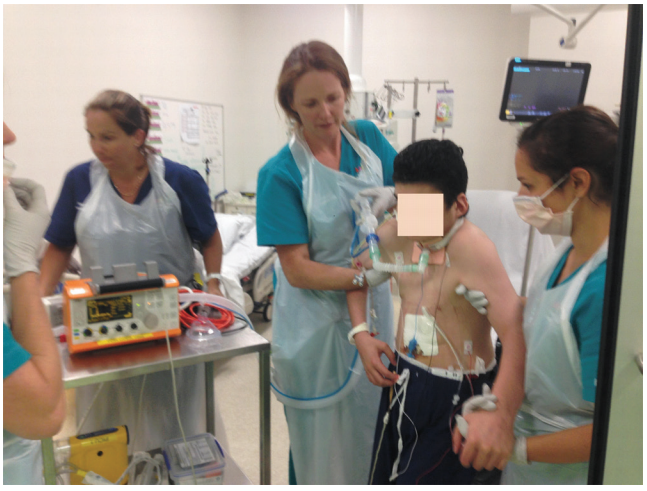

Fig. 17.30 Mobilizing a young man on the intensive care while ventilated via a tracheostomy.

intubated child, ACTs are often used in combination for greater efficacy and appropriate anticipatory strategies put in place prior to treatment (e.g., preoxygenation, sedation). In addition to manual techniques and positioning, manual hyperinflation (MHI) may be indicated when areas of focal atelectasis or retained secretions are identified, with greater expiratory to inspiratory flow bias critical to the maneuver (Fig. 17.29, Video 17.3).[98] The roles of ventilator hyperinflations (VHIs) and the intrapercussive ventilator are currently under investigation.

The focus in pediatric critical care is now directed toward early rehabilitation, even prior to extubation where possible (Fig. 17.30), as it has been shown to reduce ventilator days, length of stay and delirium and to increase muscle strength.[99] Recommendations in "at risk" adults also include the use of continuous passive motion (CPM) and electronic muscle simulation (EMS) daily (Fig. 17.31).[100] For some patients, inspiratory muscle training can facilitate weaning.[101] In the pediatric arena, there is evidence supporting a similar approach[102,103]; advocacy from the therapist is critical to overcome barriers and identify the children who would benefit.

AIRWAY AND STRUCTURAL DISORDERS

Asthma

A crucial part of physical therapy management of asthma is education of the child and parents, in particular the need for regular exercise when the child stable.[104] It is important to ensure ongoing adherence to prescribed medication (see Chapters 45 and 46). Physiotherapy includes advice on exercise that is specific to age and severity. In some children, breathing retraining using a reduced volume and/or frequency with relaxation can reduce symptoms and therefore improve quality of life. Several groups advocate specific techniques, but it is important to stress that these techniques are adjunctive to medication and are not replacement therapy. Routine airway clearance is rarely indicated in asthmatic patients, and it is important to remain aware that ACTs may exacerbate bronchospasm.

Breathing Pattern Disorders

This is a particularly challenging area. It is important that the child undergo a full assessment prior to referral for physical therapies to rule out organic disorders (e.g., croup) and neurologic disorders (e.g., Rett syndrome). A careful history should be taken to investigate when symptoms are present. If the child is symptom-free during sleep, this may help to refine the diagnosis. If the child is suffering from a psychogenic disorder or other abnormal breathing patterns (e.g., hyperventilation syndrome and sighing dyspnea), then physiologic input will have benefit to identify triggers and provide advice on how to cope with them. Breathing reeducation is essential in this group of children and adolescents. Breathing retraining, which incorporates reduced respiratory rate and/or tidal volume, is a first-line treatment for hyperventilation syndrome with or without concurrent asthma. Identification of precipitating factors (e.g., sighing) and other triggers is important.

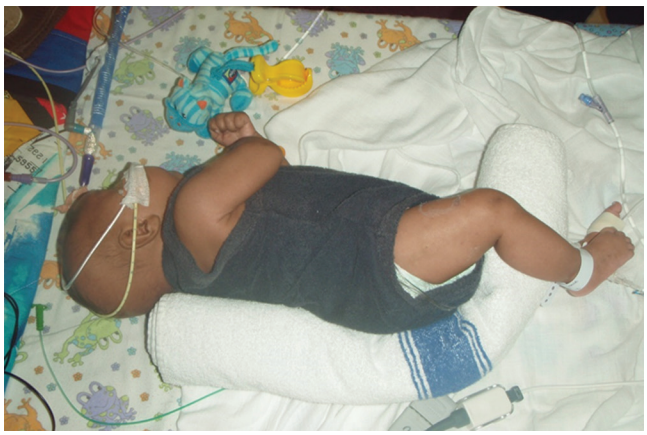

Fig. 17.32 Positioning of an infant with chronic neonatal lung disease.

Chest Wall Disorders

Physical therapies in chest wall disease are aimed at maximizing lung function. This includes pulmonary rehabilitation,[105] manual musculoskeletal therapies for postural pain, and initiation of NIV in the hypercapnic patient[106] along with postoperative rehabilitation after corrective surgery.

Airway Structural Disorders

With the increased diagnosis of tracheomalacia and bronchomalacia via bronchoscopy, physiotherapists are building evidence for the use of PEP.[107] It is hypothesized that in children with some structural abnormalities, PEP can increase airway stabilization and enhance cough expiratory flow.[108] It is well known that children with airway malacia are prone to recurrent respiratory infections,[109] and further research is needed on the role of physiotherapy.

Interstitial Lung Disease

There is little published evidence on physiotherapy for interstitial lung disease. Studies in adult patients have shown an improvement with exercise training in patients with this disease.[110–112] Adult patients with more advanced disease may benefit from ambulatory oxygen therapy and breathlessness management. Physical therapies for this condition include positioning to ease breathlessness. Given the very different spectrum of interstitial lung diseases in children (Fig. 17.32 and Section 6: Interstitial Lung Disease, of this book), recommendations based on adults should only be adopted with extreme caution and a detailed individualized assessment is essential.

Conditions Not Generally Amenable to Physiotherapy

ACUTE LARYNGOTRACHEOBRONCHITIS (CROUP)

Physical therapies are usually contraindicated in the spontaneously ventilating child with croup. However, other techniques may be indicated if the child is intubated and mechanically ventilated and secretions cannot be cleared by suction airway clearance alone.

PERTUSSIS

Any physical therapies during the acute phase of pertussis can precipitate paroxysmal cough and its complications. If the child is mechanically ventilated, paralyzed, and sedated and there are issues with retained secretions, ACTs may be of benefit (see Figs. 17.3–17.5). In the child with persistent lobar collapse and in whom the paroxysmal cough phase has ended, appropriate ACTs can be taught.

INHALED FOREIGN BODY

Physical therapy is not indicated to remove the foreign body. These children require bronchoscopic removal of the material, usually with a rigid bronchoscope. After bronchoscopic removal, PD, manual techniques, and breathing exercises may be necessary to clear excess secretions that have accumulated in the obstructed airway behind the foreign body.

PULMONARY EDEMA

ACTs are not indicated for pulmonary edema; however in some situations CPAP or NIV may be an appropriate strategy to help children with significant work of breathing while waiting for diuresis or other medical strategies to take effect.

LOBAR PNEUMONIA AND EMPYEMA

There is little evidence to support chest physiotherapy to treat lobar pneumonia[113,114] or empyema.[115] The pathophysiology of both conditions indicates that physiotherapy will not be effective in the acute stages or even during the resolution phase when secretions can appear in the airways; previously healthy children have the capability to clear these independently, with particular focus on mobilization. However, specific ACTs may be required in children with conditions that alter muscle tone (e.g., cerebral palsy), mobility/muscle strength (e.g., neuromuscular conditions), or mucociliary clearance (e.g., bronchiectasis).

BRONCHIOLITIS

Physical therapy is counterproductive during the acute stage of bronchiolitis. Mechanically ventilated infants will need careful assessment and may benefit from ACTs if there is retention of secretions. Chest physiotherapy has not been shown to reduce length of hospital stay, oxygen requirements or clinical severity score[116]; intervention should be based on specific focal signs or comorbidities. In these cases, physiotherapy may need to be modified to reduce the impact on the infant's work of breathing and should be continually reassessed.

In children with normal lung defense mechanisms and function, chest physiotherapy is very unlikely to be of benefit for acute respiratory disorders. However, lack of evidence does not mean that this should be extrapolated to all children. It is imperative that the therapist be able to assess the patient and liaise with the medical team regarding the necessity of intervention so that appropriate treatment is provided.[117]

Summary

Physical therapy is well established as part of the management of many respiratory conditions, in particular for children requiring mechanical ventilation and those with chronic disorders such as bronchiectasis or neuromuscular conditions. Over the past few decades, the profession has evolved, and a wide variety of techniques and modalities is available with a growing evidence base. Fundamentally the key to effective physiotherapy is identifying the physiologic issue, deciding whether physiotherapy strategies can assist, and identifying outcomes that can be measured. The latter must include both positive and negative effects so the therapist can assess the risk and take an informed approach to patient care. Integral to all of this are the therapeutic skills required in communication and engagement with the child and family to ensure an optimal outcome in the ever-changing face of the presentation.

Acknowledgments

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