11 Physical Therapy Following Phrenic Nerve Graft Surgery: Implications Far Beyond Breathing

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Introduction

Decreased endurance and dyspnea may be the primary presenting symptoms of unilateral or bilateral diaphragm paralysis, but further evaluation may expose a host of other adverse physical consequences. In order to understand the wide array of physical symptoms reported with unilateral or bilateral diaphragm paralysis, it is critical to understand the diaphragm's role as the body's major pressure regulator.

The diaphragm plays a major role in dynamic stabilization of the trunk and spine [1, 2]. The diaphragm is situated in the middle of the trunk, completely separating the thoracic and abdominal cavities, with external valves at the top (vocal folds) and at the bottom (pelvic floor). Hence, the diaphragm's movements cause a constant fluctuation in intra-abdominal and intrathoracic pressures, allowing the diaphragm to simultaneously manage respiration and spinal stabilization needs [3, 4]. This can be easily visualized as a soda pop can which uses internal pressure to stabilize the flimsy aluminum walls (dynamic spinal stability) (see Fig. 11.1) [5]. The ability of the trunk to control multiple functions, such as respiration, spinal stability, balance, limb force production, voicing, and continence, is dependent on finely regulated pressure changes, with the diaphragm as the prime contributor [6–8]. Thus, with diaphragm paralysis, besides the obvious breathing impairment, a whole range of unintended consequences need to be assessed.

When disease or trauma, such as a phrenic nerve injury, prevents or limits the diaphragm from significantly contributing to inspiration, there is a substantial risk of secondary problems with balance and spinal control [9, 10]. The altered motor control of the trunk can lead to musculoskeletal consequences including chronic pain (most often low back, neck, shoulder, or hip pain), shoulder range of motion restrictions, and/or pelvic floor dysfunction [8, 11]. The relationship of the diaphragm to upright postural control goes far beyond inspiration. The critical relationship of the diaphragm to respiratory and postural control function means that all patients with phrenic nerve paralysis should be screened for impairments of both functions.

Physical Therapy (PT) Preoperative Assessment

A physical therapist should assess the patient's breathing and postural presentation with this broader understanding of the diaphragm's multiple roles. Recommended assessments are listed in Table 11.1.

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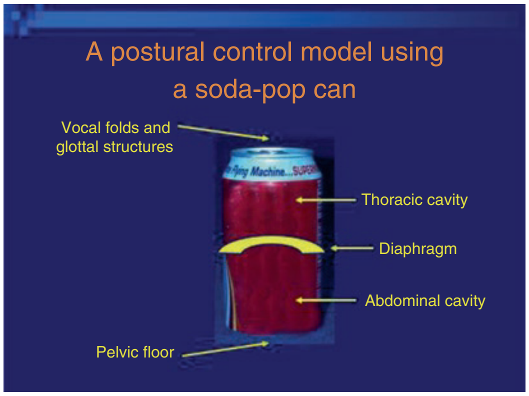

Fig. 11.1 Soda pop can model of postural control. The diaphragm is the body's major pressure regulator, completely separating the thoracic and abdominal chambers. Together with the superior valve (vocal folds) and inferior valve (pelvic floor), the diaphragm simultaneously controls trunk pressure for breathing and postural stability, which functionally links the *top* and the *bottom* of the can

Table 11.1 Preoperative PT assessment should ideally address the following potential impairments

Respiration	Pulmonary function tests Compensatory breathing patterns, assessed in multiple postures and activities Chest wall excursion (CWE) Sleep-disordered breathing
Endurance	Mobility tests (i.e., 6-min walk test) Perceived exertion during activities
Postural alignment in upright, especially	Spine and neck Rib cage Shoulders Pelvis/hips Pediatric vs. adults
Postural stability	Balance Gait deviations Pain Continence

Respiration

Pulmonary Function Tests (PFT) The combination of forced vital capacity (FVC) and the ratio of forced expiratory volume in one second to FVC (FEV1/FVC) are commonly used to clarify the severity of the lung restriction [12].

Compensatory Breathing Patterns Analysis of the patient's breathing pattern should be screened to identify which substitution patterns are used to achieve inspiratory lung volumes at rest (tidal volume) and with effort (vital capacity). Did the patient compensate in a functional manner or did the adaptive strategy add a burden to their pulmonary efficiency or postural control? PTs should assess the patient's breathing strategy in multiple postures and activities presurgically for post-nerve regeneration comparison. Specific manual palpation exam of both hemidiaphragms during inhalation should be done to determine the contributing function of each hemidiaphragm for postsurgical comparison. Where available, a videofluoroscopy or an ultrasound test can confirm or refute the therapist's manual palpation findings, which will be helpful for postoperative comparison [13, 14].

Chest Wall Excursion (CWE) CWE measurements can quantify the chest movements and have been shown to have good inter-/intra-tester reliability after minimal training [15, 16]. Suggested measurement sites that capture the common variations of breathing patterns are (1) level of the third rib (axilla), (2) xiphoid process, and (3) half the distance from xiphoid to umbilicus [17] (Fig. 11.2). CWE should be tracked through rehabilitation, hopefully showing a shift downward (increased diaphragm excursion and decreased upper chest accessory muscle recruitment), and a more symmetrical response.

Sleep Breathing assessment should encompass the entire day, thus including sleep. Bilateral diaphragm paralysis will require nocturnal support such as CPAP (continuous positive airway pressure), BPAP (bi-level positive airway pressure), or mechanical ventilation, but mounting evidence suggests that unilateral diaphragm paralysis results in sleep-disordered breathing, cespecially during REM sleep, that often goes undetected [18–21]. Sleep studies should be a routine. PTs' expertise in positioning may help patients and their physicians determine optimal sleep postures for oxygenation.

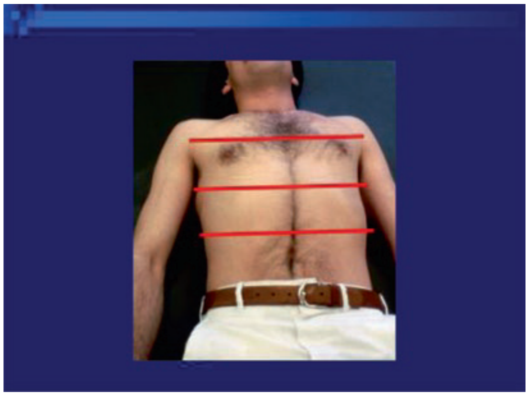

Fig. 11.2 Chest wall excursion (CWE) circumferential measurement sites: *Top line*: level of the third rib (axilla). *Middle line*: xiphoid process. *Bottom line*: half the distance between the xiphoid process and the umbilicus

Endurance

Endurance tests must be individually chosen to ensure that the measure will be sensitive enough to capture a change postsurgically. A 6- or 12-min walk test is easy to administer clinically. The length of the test is determined by the patient's level of impairment. A patient who is very weak may not be able to walk even 6 full minutes, whereas a patient with lesser impairment won't show an endurance limitation until pushed a greater distance, like the full 12-min test [22]. There are many tests to choose from that are appropriate to the patient's activity level [23].

Endurance could also be measured in terms of perceived exertion [24]. These subjective tests are particularly sensitive measures for patients who are very weak and may not show an increase in functional endurance such as walking further, but who may report less perceived effort for the same breathing and/or ADL skills recorded preoperatively.

Postural Alignment

Compensatory breathing patterns and the secondary impairments in balance may cause postural abnormalities [9]. This is particularly true for long-standing phrenic nerve paralysis where disuse has caused decreased chest wall and spinal mobility. Postural compensations may have developed slowly, and the patient may not even be aware of how much his posture has changed. Manual assessment of the entire rib cage and spine is necessary as restrictions are common in long-standing phrenic nerve paralysis. Shoulder range of motion (ROM) is often restricted secondary to diaphragm dysfunction. The weakness/paralysis disrupts the normal coupling effect between the shoulder and rib cage, especially when reaching above 90° of shoulder flexion [25, 26]. Limitations may also be noted at the hip/pelvis or neck due to compensatory breathing and/or postural control strategies. PTs should anticipate the need to mobilize noted musculoskeletal restrictions in order to regain maximal breathing and postural control function.

Postural impairments secondary to unilateral diaphragm paralysis are unique. These patients should be specifically screened for asymmetrical presentations. The development of chronic pain from asymmetric muscle use is a common complaint in this population but not yet researched (Fig. 11.3). Recent research suggests that patients with unilateral diaphragm paralysis have increased balance impairments because their center of mass is disturbed with every breath on a coronal plane, rather than just a sagittal plane [9]. Compensations to correct for this added postural burden could lead to repetitive muscle stress resulting in pain.

Pediatrics Special consideration must be made in pediatrics as their chest wall and spine have not yet matured. Chronic phrenic nerve paralysis may lead to hypoplasia of the rib cage on the side of the paralysis which in turn will contribute to scoliosis forces and increased likelihood of more pronounced asymmetric postural alignment throughout the trunk (Fig. 11.4). Balance compensations will increase the risk of repetitive stress injuries as the child matures, which could lead to pain. Recent research shows that patients with chronic low back pain have weaker diaphragms than controls [27]. Long-term research has not yet been done with children with pediatric

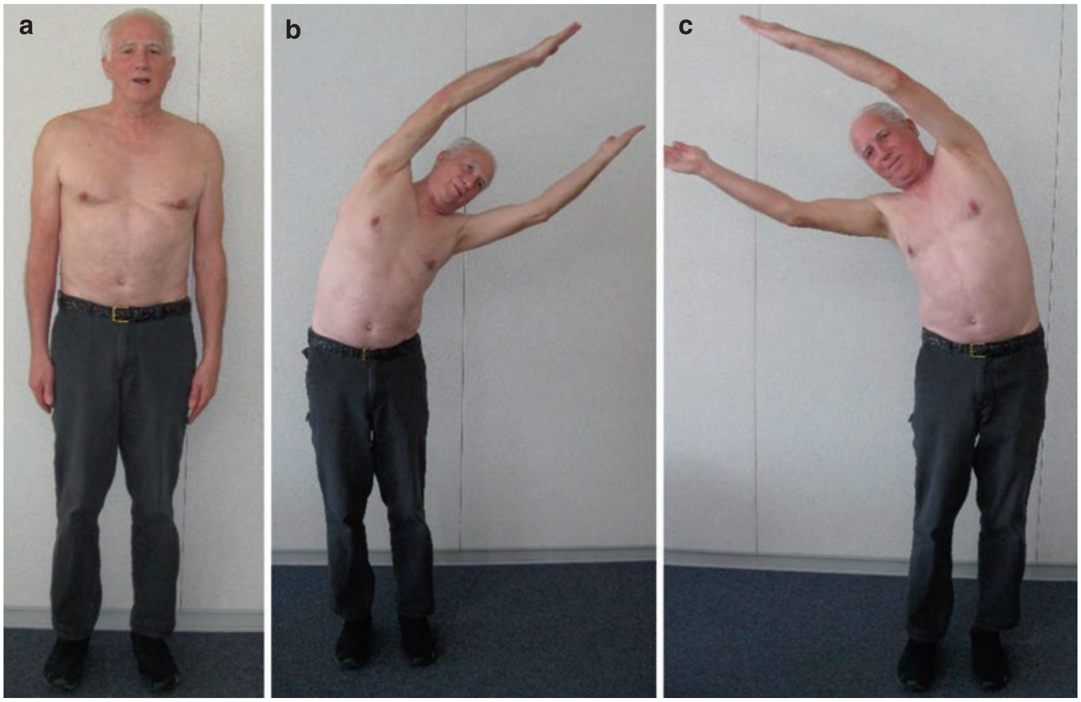

Fig. 11.3 Common postural compensations/abnormalities with unilateral phrenic nerve injury. Previously healthy, fit 67-year-old man with left paralyzed phrenic nerve of >12-month duration. Phrenic nerve graft surgery ~4 months prior to these pictures. One PT visit thus far. (**a**) Subject's trunk is shortened on the left. He reported no postural asymmetry problems before phrenic nerve injury. (**b**) Lateral side bending to left shows normal range of motion for the right rib cage and lower trunk (intact phrenic nerve). Patient shows normal weight shift of hips to the right when side bending to left. (**c**) Lateral side bending to the right shows moderate restrictions in left lower rib cage and lower trunk (left paralyzed phrenic nerve). Note: (1) increased effort in his face, (2) decreased elbow extension, (3) decreased shoulder flexion, and (4) decreased weight shift onto left lower extremity. These restrictions limit more than inspiratory lung volumes, such as limitations in balance, gait, and reach, and may lead to chronic pain from long-standing altered mechanics

phrenic nerve injuries but would suggest that maturing with a chronically weak or paralyzed diaphragm would predispose these children to an increased risk of developing low back pain.

Postural Stability

There are numerous functional presentations of postural instability. Four specific impairments are detailed below.

Balance The contributions of the diaphragm to postural stability are well established; thus, the assessment of balance (postural instability) should be a routine screening module of the PT evaluation. There are numerous balance tests from simple sitting perturbation tests, to timed single-limb stance tests, to sophisticated computerized limits of stability tests. Balance tests should be chosen for their sensitivity to change, availability of resources, and specificity to each individual patient's capabilities [28].

Gait Postural control impairments may be reflected as abnormal gait patterns and are highly variable with this population. Patients with mild postural instability are likely to stiffen their trunks in an attempt to improve postural stability. Common gait deviations associated with rigid trunks include decreased arm swing, decreased trunk rotation, slower cadence, and shorter steps. This pattern is very similar to other physical impairments such as chronic low back pain, neuromuscular weakness (Parkinson's, multiple sclerosis, stroke, etc.), incontinence, and lower

Fig. 11.4 Pediatric postural impairments 1–1/2-year-old girl surviving a traumatic vaginal birth with resultant bilateral phrenic nerve injury (left more impaired than right). Note postural shift of her trunk to left side (increased weight bearing on left leg) and increased lateral trunk flexion to left (increased skin fold), both likely balance compensations. Long term, this posturing will lead to greater scoliotic forces on her developing spine and greater risk of repetitive stress injuries (pain)

gastrointestinal dysfunctions (constipation, irritability, etc.) [29–31]. Careful screening is needed to ascertain gait deviations due to phrenic nerve paralysis vs. other underlying problems.

Patients with severe postural instability can't stiffen their trunk because of extreme weakness or motor control issues. Their trunk is too floppy, and their gait pattern typically shows deviations such as excessive arm swing and excessive trunk movements, especially in the coronal plane. If the trunk is markedly unbalanced, the patient may require an assistive device or may no longer be ambulatory.

Pain

A possible long-term consequence of altered motor plans is pain due to malalignment or overuse syndromes [32, 33]. Pain conditions may reflect overuse of the accessory muscles due to the diaphragm weakness (neck or chest pain) or due to postural instability (pain anywhere from neck to hips) [34]. Patients with unilateral diaphragm paralysis may have pain related to rotational (torsion) forces due to asymmetric and rotary forces across their spine. Compensatory trunk postural strategies could result in pain down in the knee or ankle joints. All patients should be screened for secondary pain conditions.

Continence

The deep abdominal muscle shell that controls intra-abdominal pressure is comprised of the diaphragm as the top dome, the transversus abdominis and multifidus as the long cylinder, and the pelvic floor as the bottom sling. When one of those four muscles is impaired, it impairs the function of the whole abdominal complex. Thus, diaphragm dysfunction is highly associated with pelvic floor consequences, such as incontinence, and should be screened in this population [6, 8].

Summary

Diaphragm paralysis has marked implications for motor dysfunction beyond breathing and endurance impairments and should be carefully assessed and documented preoperatively in order to accurately assess the long-term outcomes of the phrenic nerve graft surgery. However, because this surgery is so specialized, requiring many patients to travel a great distance, there is little likelihood that the same PT will do the pre- and post-surgery which may limit the reliability of the measures. Research is needed to determine the optimal pre-/posttests that could be performed reliably on a nationwide basis.

Physical Therapy Rehabilitation

PT Reassessment

Following phrenic nerve graft surgery and after the confirmation from the surgeon that it is safe to start rehabilitation, the patient should be reassessed by PT. If the patient was not seen by PT preoperatively, the PT evaluation described earlier in the chapter should be done. Two specific tests should be included in the postsurgical rehabilitation phase of PT: (1) integument restrictions and (2) diaphragm responsiveness.

Integument Surgical scars on the calf and chest, as well as surrounding tissue, should be evaluated for fascial restrictions or scar adhesions which can limit mobility [35]. If restricted, the PT should include myofascial releases and scar massage to affected areas to maximize range of motion and ease of movement.

Diaphragm Each hemidiaphragm should be reassessed with manual facilitation to determine if the diaphragm is now contracting. This is a clinical examination. Where possible, the results should be compared with a videofluoroscopy or an ultrasound of the diaphragm for confirmation [13, 14].

PT Rehabilitation Treatment

PTs are very familiar with designing treatments to address issues such as impaired endurance, poor posture, poor core stability, impaired balance, fascial/scar restrictions, gait deviations, pain, and stress incontinence. PT treatments uniquely related to phrenic nerve rehabilitation will be specifically addressed below: (1) thoracic spine and rib cage musculoskeletal restrictions, as well as (2) breathing neuromotor retraining.

Thoracic Spine and Rib Cage Musculoskeletal Restrictions Restrictions of the spine and rib cage are very common in this population due to disuse and atrophy over a prolonged period of time. Rehabilitation often includes (1) spinal mobilization techniques in the sagittal and transverse planes, sometimes in the coronal plane, and (2) rib mobilizations. The PT will need to determine if the entire rib cage or only one side (unilateral phrenic nerve injuries) needs musculoskeletal interventions. Typically, treating these trunk restrictions includes a combination of joint mobilizations, soft tissue releases, and fascial releases in order to maximize functional gains following the regeneration of the phrenic nerve [36]. Compensatory breathing and/or postural control strategies may lead to additional musculoskeletal problems which should be addressed on an individual basis.

Breathing Neuromotor Retraining Significant neuromotor retraining is necessary to stimulate and strengthen the diaphragm's response both unilaterally and bilaterally. The diaphragm also needs to be coupled with the intercostals to optimize inspiratory lung volumes [37, 38]. Adequate intra-abdominal pressure to optimize the diaphragm's length-tension relationship is imperative, so focusing on core strength is a hallmark of a diaphragm rehab program [39]. If the patient is too weak to generate adequate intra-abdominal pressures on their own, then an abdominal binder should be trialed [40].

Reduce Postural Demands on the Diaphragm Reducing the postural demand on the diaphragm will allow the diaphragm to focus on its respiratory role rather than its postural role. Thus, early in rehabilitation, facilitation of the diaphragm in recumbent postures (less postural demand) is usually more successful than upright postures. Manual facilitation techniques based on the neurophysiologic response of the diaphragm as well as the physiologic need to breathe (survival response) will bolster the clinician's efforts to elicit a response from the recovering diaphragm [41] (Fig. 11.5). Long term, the goal is to restore the diaphragm's ability to function simultaneously as a breathing and a postural control muscle; thus, recruitment of the diaphragm in higher-level postures, such as sitting, standing, walking, and running, is an important progression of treatment. Combining diaphragm breathing with complex movements including upper extremity reaching (coupling the diaphragm and intercostals) and/or gait (coupling the diaphragm with the abdominals and pelvic floor) will promote restoration of the diaphragm to its previous complex motor functions (Fig. 11.6). Resisted movements, such as with proprioceptive neuromuscular facilitation (PNF) exercises, will drive a greater motor response

Fig. 11.5 Neuromotor retraining: handling techniques to maximize the response of a weak diaphragm contraction. (**a**) "Diaphragm scoop" neuromotor facilitation technique can be used as a diagnostic screening tool to assess bilateral or unilateral diaphragm contraction and used to stimulate the diaphragm's contraction. Resistance is added once the diaphragm is consistently activating. Rehabilitation starts with isolated diaphragm facilitation techniques but quickly moves to integrated complex neuromotor retraining that includes coordinating simultaneous postural control responses of the diaphragm with the breathing responses of the diaphragm (ventilatory strategies). (**b**) "Lateral costal" neuromotor facilitation technique progresses from a focus on facilitation of a central response of the diaphragm (hemidiaphragm or bilateral) to a coupled response with the intercostals (progression toward normal multipurpose reaction)

Fig. 11.6 Sample of techniques that promote integration of diaphragm with higher-level postural stability such as standing, reaching, and axial trunk rotation. (**a**) Mid-trunk dynamic stabilization resistance training in standing requires the diaphragm to move dynamically for breathing as well as postural stability (balance). The therapist applies resistance at the mid-trunk to actively engage the diaphragm in balance response. Submaximal resistance allows breathing and postural control responses. Avoid maximal resistance as it will elicit a static breath-hold response and is not a desired outcome. (**b**) Distal resistance shown here on the arm, but can also be done on the leg, demands that the diaphragm respond as a postural muscle. Be careful to use submaximal resistance so that the diaphragm can relearn how to breathe and "hold" at the same time (dynamic stability). (**c**) Rotary resistance of the trunk, shown here in sitting but can be done in any upright posture, focuses on the role of the diaphragm for balance responses (maintenance of axial control of the trunk in upright)

from the diaphragm and trunk muscles which will aid in strengthening the reemerging complex neuromotor plans [42–44].

Unilateral Paralysis Diaphragm Neuromotor Retraining Specific neuromotor retraining techniques that bolster the response of the weak side of the diaphragm are particularly helpful. This author highly recommends using proprioceptive neuromuscular facilitation (PNF) "timing for emphasis" technique to use the strength of the strong diaphragm to get an overflow response to the weak hemidiaphragm [41, 45]. Immediately after eliciting the motor response, the diaphragm's efforts should be reinforced by pairing inspiration with activities that naturally couple the diaphragm with the intercostals and abdominals such as reaching or trunk rotation. This will aid in reinforcing the diaphragm's inspiratory response and promote the diaphragm's role as a postural stabilizer as well.

Endurance Training for the Diaphragm Once the diaphragm responds consistently, ventilatory muscle training programs (inspiratory and expiratory muscle trainers) can be initiated to increase endurance and reinforce neuroplasticity [46]. Don't start these programs until the diaphragm has adequate strength or the other inspiratory muscles may overpower the diaphragm's response, which would have the unintended outcome of reinforcing the compensatory pattern instead of strengthening the diaphragm.

Airway Clearance If the patient has a history of decreased ability to clear lung secretions secondary to diaphragm weakness or paralysis, then a comprehensive airway clearance program should be developed, including assistive cough techniques [47].

Summary

Rehabilitation of the diaphragm following a phrenic nerve graft involves a detailed multisystem evaluation by the PT to determine the extent of the impairment (primary and secondary) as a result of the long-standing unilateral or bilateral phrenic nerve paralysis. A bilateral paralysis has a devastating impact on the patient's survival, often requiring mechanical ventilation, but unilateral paralysis is also devastating due to balance impairments, endurance impairments, sleep disruptions, musculoskeletal restrictions/pain, and an ongoing risk of respiratory complications. If the consequences of phrenic nerve paralysis are not fully understood, assessed, and treated, the patient's long-term quality of life outcomes may be impaired.

Dr. Kaufman's surgical approach to phrenic nerve paralysis is an innovative procedure. Research with spinal cord injury gives PT's guidance for the development of an appropriate treatment approach to this population, but research specific to phrenic nerve restoration is needed.

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