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Duchenne and Paradoxical Respiration

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Background

As noted in prior historical vignettes, in the early 1900s there was reasonable understanding of the brainstem's respiratory center. Its three components of neural mechanisms (the central and peripheral chemosensitivity, central respiratory drive, and neuronal feedback from the muscles of respiration) were identified and characterized in the late 1800s and early 1900s (The colorful neurologist Brown-Séquard disputed whether the spinal cord had respiratory centers as well but was forcefully rebuffed [1–3]).

But drive is not enough; force and motion are needed. Similarly, investigators became better acquainted with the function of the respiratory muscles and its neural connection with cranial nerves (CN): e.g., the muscles moving the alae nasi to decrease airway resistance (CN VII), the muscles opening the glottis (CN X), the major inspiration diaphragm muscle (cervical cord by the third to fifth cervical nerves merging into the phrenic nerve), and intercostal muscles from the dorsal spinal cord needed as backup during dyspnea. The neurology of respiration became divided into central and peripheral. Traditionally, "peripheral" implied the trajectory of the phrenic nucleus to the muscle fibers of the diaphragm.

The history of lung mechanics started before there was even a basic idea of central nervous system regulation. For example, Galen (Claudius Galenus AD 131–201) in *On the Natural Faculties* was unclear and conflicted on the purpose of breathing (considered by some to cool inborn heat), but he identified the diaphragm as a major muscle not only separating the chest from the abdomen but also as a muscle of respiration [4]. Galen's most important books on respiration, *On the Causes of Respiration* and *On the Movements of the Thorax and Lung*, have been lost, and history can only look back at what was subsequently cited by later researchers [4, 5]. *On the Causes of Respiration* describes Galen's original findings about chest wall movement, including the discovery of two layers of intercostal muscles and the enumeration of all respiratory muscles with demonstration of their innervation. Galen also applied what he learned about the diaphragm in real life. He emphasized observing the motion of the chest wall and noting which respiratory muscles were used and which ones failed when he severed the spinal cord in pigs.

The great anatomist, physiologist, and illustrator, Leonardo da Vinci, was seriously interested in lung mechanics and lung inflation, and his drawings showed some preliminary concept of nervous control and the biomechanics of muscles [6]. No one followed up these early observations with new research for centuries. The anatomists of antiquity recognized only one single respiratory muscle that could raise and carry the ribs and did not look beyond the diaphragm.

Since the early 1800s, biologists have recognized that the cavity of the thorax enlarges in all directions during inspiration. The lungs expand to fill the created space. When the muscles relax, the gravitational force and elastic reaction of the thoracic wall and pulmonary tissue bring the thorax back to its original size. All mammals breathe with this mechanism; inspiration requires muscle contraction, and expiration is largely passive.

In the late 1800s, the physiologist Henry Newell Martin refocused on the muscles of respiration; he attempted to elucidate the additional role of the internal intercostal muscles and their function as accessories when the diaphragm fails and during extreme dyspnea. "*The diaphragm, when the apnea passed off, made a few contractions without any activity of the intercostal muscle: but this latter soon began to contract in regular alternation with the diaphragm and before the occurrence of expiratory convulsions; in fact with the commencement of dyspnea*" [7]. These muscles do have an inspiratory function but a secondary one, and over

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time, investigators also found an inspiratory function for the sternocleidomastoid muscles, which become active during labored breathing. Shäfer's *Text Book of Physiology* in 1900 clearly identified them without specific attribution but also mentioned abdominal muscles for forced expiration. These basic biomechanics have remained the foundation of our understanding of respiratory mechanics [8] (Fig. 1).

A number of observations were necessary to explain the coordinated mechanics. First, the lungs are elastic, and the elastic fibers allow distention of the lung with distending force, but they easily collapse if this distending force is removed. We know that piercing the thoracic cage equalizes pressures on the outer and inner walls of the lung, and the lung collapses, causing air to get sucked in from the hole. Physiologists thus found the lung in a state of tension with pull on the thoracic wall, creating a negative pressure in the pleural cavity. This negative pressure increases with inspiration and decreases with expiration. Inspiration causes elevation of the ribs and descent of the diaphragm. Raising the ribs straightens the angle between rib and costal cartilage, pushing the sternum forward and upward. When the muscular fibers of the diaphragm contract, the diaphragm lowers and flattens its circumference. The central tendon of the diaphragm is motionless. When labored respiration occurs, other muscles, including the sternomastoid, the pectoralis minor, the lower part of the pectoralis major, and the lower slips of the serratus magnus, take over. Investigators developed and viewed pneumatographs and phrenographs (when he was a medical student looking at lung physiology and not lung mechanics, neurologist Henry Head created a pneumograph that produced the most reliable results).

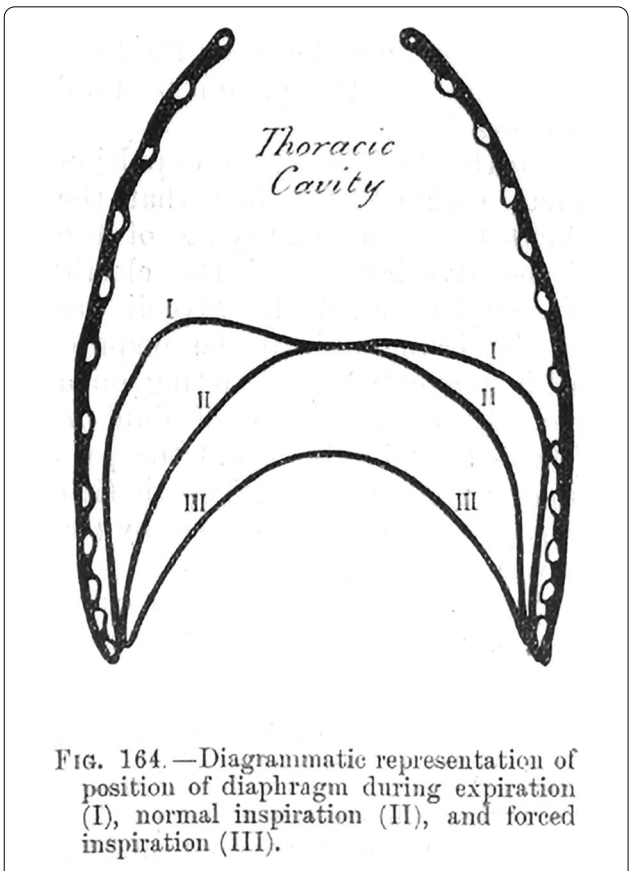


Fig. 164.—Diagrammatic representation of position of diaphragm during expiration (I), normal inspiration (II), and forced inspiration (III).

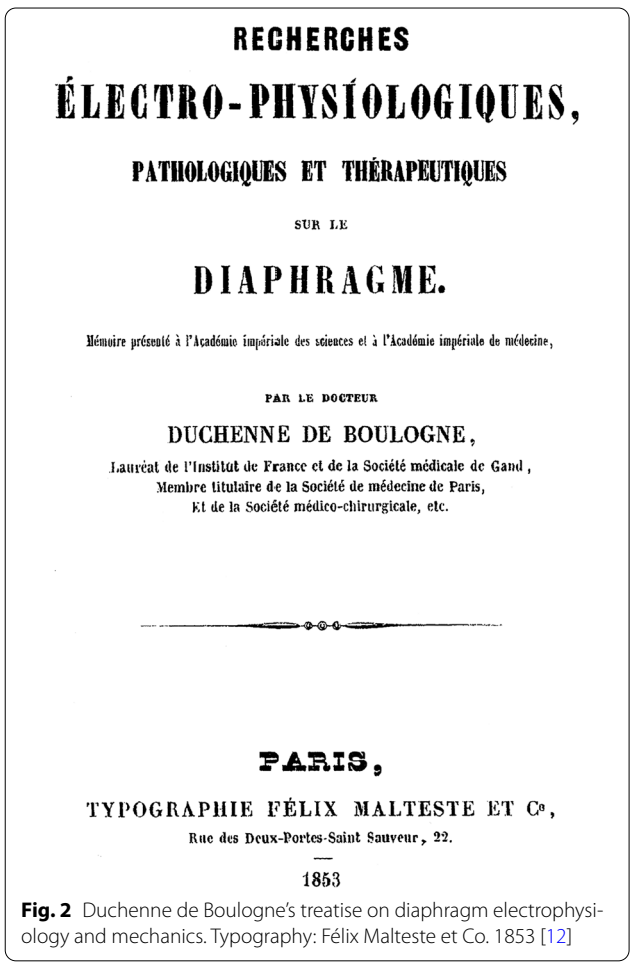
Fig. 1 Illustration of diaphragmatic movement (from Shäfer EA. *Textbook of Physiology* Young J Pentland Edinburgh & London 1900 [8])

Later developments included the identification of the phrenic nerve motor neurons in the midcervical (C3–C5) spinal cord, "a straight, very discrete column of cells exactly paralleling the ventral longitudinal fissure)" [9]. The phrenic nerve is a mixed sensory-motor nerve, and only recently, investigators discovered that sensory-afferent neurons project to the cortex, explaining the perception of air hunger by patients. However, the focus rapidly shifted back to characterization of the central respiratory drive, and interest in the workings of respiratory muscles waned. Muscle investigations were performed in the nineteenth century by Beau and Maissiat [10], Bert [11], and Duchenne de Boulogne [12]. Messieurs Beau and Maissiat concluded from their experiments that the diaphragm, by its own action, elevates the diaphragmatic ribs.

Duchenne and Lung Mechanics

Duchenne's contribution to respiratory mechanics is most notable and presented here for the first time. Known as the Salpetrière neurologist, he described a muscular dystrophy (DMD), which we now know is a genetic muscle-wasting disease and the most commonly inherited pediatric myopathy. Advanced therapeutics in pulmonary care have significantly reduced respiratory complication-related mortality, making cardiomyopathy the main determinant of survival. Ventilatory failure from muscle weakness requires mechanical support for ventilation as soon as there is symptomatic nocturnal hypoventilation. Today, noninvasive positive-pressure ventilation is the method of choice for supportive long-term mechanical ventilation in DMD. With the progression of DMD, the diaphragm becomes progressively weak, and its action reduces to the point that paradoxical cephalic movement may occur while its thickness increases, indicating pseudohypertrophy due to infiltration of connective tissue and fat deposition. The diaphragm is prone to pseudohypertrophy in the youngest patients with DMD and to progressive atrophy in middle-aged and older patients.

Virtually forgotten is Duchenne's essay on lung muscle mechanics (Duchenne was also known for his faradic-stimulation studies, which made us understand facial changes with emotions.) In his article on lung muscle mechanics [12] (Fig. 2), Duchenne demonstrated the movements of the ribs by the diaphragm and how muscle retained its natural relationships with the abdominal viscera. He found that excitation of the phrenic nerve only produced contraction of the diaphragm. He also concluded that paralysis of the diaphragm is not fatal in itself, as was generally believed ("*La paralysie du diaphragme n'est pas en elle-même mortelle, comme on le pense généralement*"). He found that the action of the intercostals drove inspiration when the patient was at rest. "The patient, in fact, lives a long time with paralysis of the diaphragm, but then the simplest bronchitis can cause death by asphyxiation, expectoration being difficult or impossible" ("*Le malade, en effet, vit longtemps avec une paralysie du diaphragme; mais alors la plus simple bronchite peut occasionner la mort par asphyxie, l'expectoration étant difficile ou impossible*".)


RECHERCHES

ÉLECTRO-PHYSIOLOGIQUES,

PATHOLOGIQUES ET THÉRAPEUTIQUES

SUR LE

DIAPHRAGME.

Mémoire présenté à l'Académie impériale des sciences et à l'Académie impériale de médecine,

PAR LE DOCTEUR

DUCHENNE DE BOULOGNE,

Lauréat de l'Institut de France et de la Société médicale de Gand,
Membre titulaire de la Société de médecine de Paris,
Et de la Société médico-chirurgicale, etc.

PARIS,

TYPOGRAPHIE FÉLIX MALTESTE ET Cᵉ,

Rue des Deux-Portes-Saint Sauveur, 22.

1853

Fig. 2 Duchenne de Boulogne's treatise on diaphragm electrophysiology and mechanics. Typography: Félix Malteste et Co. 1853 [12]

He was arguably the first to recognize paradoxical breathing. "At the time of inspiration, the epigastrium and the hypochondria become depressed while the chest expands; the movements are in an opposite direction, during expiration." ("*Au moment de l'inspiration, l'épigastre et les hypocondres se dépriment pendant que la poitrine se dilate; les mouvements de ces mêmes parties se font dans un sens opposé, pendant l'expiration.*") He continued, "This would not bother the patient too much, but if the patient comes to make some effort, either to walk or to talk, [such] that he experiences the slightest impression, his breathing accelerates (48 or 50 inspirations per minute); the trapezius, sternomastoid, serratus, pectoralis major, and latissimus dorsi muscles enter into contraction; his face reddens, [and] the patient suffocates. He is forced, if he walks, to sit down after a few steps, or if he wants to speak, to take breath to continue his sentence, which he cannot finish without stopping every moment." He concluded that the contraction of the diaphragm changes the anteroposterior diameter of the base of the thorax but in a barely perceptible way. When an animal is disemboweled and the viscera has been lowered, the isolated contraction of the diaphragm, produced by localized electrification, carries the diaphragmatic ribs in an opposite direction, that is to say, inward.

Further understanding of neuromuscular respiratory failure came with better understanding of the pathophysiology in acute neuromuscular disease, and clinicians led the way in explaining how diaphragm function and the upper airway could be affected. The rapid admission of a large number of dyspneic patients during the poliomyelitis epidemics put neuromuscular respiratory failure front and center. Many early neurologists were interested in this acute neurologic disease, and poliomyelitis was perhaps the most important and most threatening disorder appearing in sudden epidemics in the early 1950s.

We cannot be certain if Duchenne was the first neurologist to become interested in the neuromechanics of respiration, but his 70-page treatise was a major contribution. Contemplating the role of neurologists in pulmonary mechanics, we already came across work from the United States and United Kingdom. The details of airway and respiratory recognition and suggestions for the most appropriate management reported by Russell, Baker, and Plum are also major contributions [13]. It is a common misunderstanding that neurologists entered this arena after other specialties, such as anesthesia, defined critical care medicine. The seeds of interest in (what later became) neurocritical care may have been planted much earlier than appreciated.

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Conflict of interest
There are no conflicts of interest.

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