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Landmark and Legacy

# Congenital diaphragmatic hernia: lessons learned and lost

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ABSTRACT



**Contents**



The story of congenital diaphragmatic hernia illustrates the role of past legacies and 20th century landmarks in our understanding of this complex disease. It is also a tale of lessons learned and lost.John Baptist Morgagni described infants with intestine herniated into the pleural spaces, and an adult who lived to a ripe old age with a substernal opening, the Foramen of Morgagni. The early anatomists considered these infants to be anatomic curiosities or 'monsters' [1,2]. Sir Astley Cooper, a surgeon to Kings, described two infants with 'unnatural' openings between their chests and abdomen, and said the hypoplastic lung was the size of a 'nutmeg' [3]. (See Figs. 1–3.)

Henry Bowditch, a colleague of Oliver Wendell Holmes, diagnosed a congenital diaphragmatic hernia in a seventeen-year-old laborer when he heard gurgling intestinal sounds over the left chest. The patient said his heart had always been on the right side. The young man died from an injury, and at autopsy, the right diaphragm was absent. The stomach and most of the intestine were in the left thoracic cavity. Dr. Bowditch said it was "evidently a foetal arrest of development". He made the diagnosis in 1846, and in 1853 published the case and reviewed the world's literature [4].

In 1848, Vincenz Alexander Bochdalek, a professor of anatomy in Prague, meticulously dissected the pleuro-peritoneal canal and recognized the increased incidence of left sided hernias [5]. His work gave rise to the terms, 'Foramen of Bochdalek' and 'Bochdalek Hernia.' He recommended 'practical surgery' as the only cure.

In 1889, Joseph O'Dwyer, a surgeon at the New York Foundling Hospital, operated on a three-and-a-half-year old child who was thought to have an empyema. Instead, he found intestine in the chest, which he pushed into the abdomen and sutured a hole in the diaphragm. The child died six hours later. He blamed the death on increased intra-abdominal pressure because the intestine had lost its 'right of domain' [6]. This observation on increased intra-abdominal pressure is as important today as in 1889. Dr. O'Dwyer invented an endotracheal tube to treat laryngeal obstruction in diphtheria and died after contracting diphtheria from a patient.

The discovery of roentgen rays led to the diagnosis and surgery in more children during the early 20th century. There was often no clear differentiation between congenital and traumatic hernias. Many of the children with chronic coughs, cyanosis, and malnutrition were diagnosed with tuberculosis. During this period, all diagnosed in infancy died [7,8].

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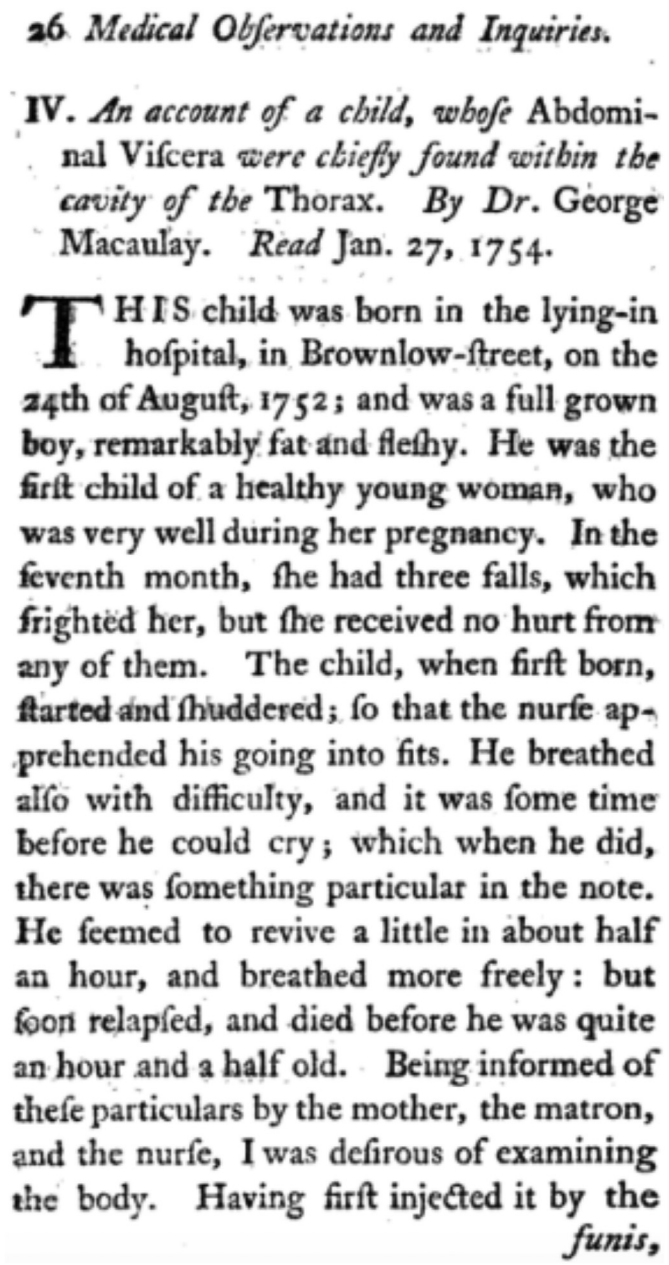

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IV. *An account of a child, whoſe* Abdominal Viſcera *were chiefly found within the cavity of the* Thorax. *By Dr.* George Macaulay. *Read* Jan. 27, 1754.

THIS child was born in the lying-in hoſpital, in Brownlow-ſtreet, on the 24th of Auguſt, 1752; and was a full grown boy, remarkably fat and fleſhy. He was the firſt child of a healthy young woman, who was very well during her pregnancy. In the ſeventh month, ſhe had three falls, which frighted her, but ſhe received no hurt from any of them. The child, when firſt born, ſtarted and ſhuddered; ſo that the nurſe apprehended his going into fits. He breathed alſo with difficulty, and it was ſome time before he could cry; which when he did, there was ſomething particular in the note. He ſeemed to revive a little in about half an hour, and breathed more freely: but ſoon relapſed, and died before he was quite an hour and a half old. Being informed of theſe particulars by the mother, the matron, and the nurſe, I was deſirous of examining the body. Having firſt injected it by the *funis*,

**Fig. 1.** Title page from Macaulay's original account of a newborn infant born with a congenital diaphragmatic hernia. From ref. no. [2].

Philomen E. Truesdale, in Fall River, Massachusetts, had a special interest in children with diaphragmatic hernia. Other physicians referred "hopeless" patients to him, and in 1935 he reported thirteen operations in children ages one to thirteen years with congenital and traumatic hernias. Only one died, who was a nine-year-old boy with a pectus excavatum and rheumatic heart disease [9].

Dr. Henry Johnson, in Glendale, California, operated on a newborn infant who had cyanosis and dyspnea at birth. X-rays demonstrated intestine in the left chest. Dr. Johnson made an abdominal incision, reduced the viscera, and closed the diaphragm with catgut sutures. The baby took breast milk four hours after the operation and was thriving at one year of age [10]. Late in the same decade, one thirteen-day old baby and a three- and a half month old infant, both in Chicago, survived surgery [11,12]. This is remarkable since they were operated upon under drop ether anesthesia without antibiotics, mechanical ventilation, blood transfusions, Intravenous fluids, or infant incubators.

The most important 'legends' in this saga are William E. Ladd and Robert Gross whose great skill and insight led to successes in infants as young as 24 h of age. Dr. Gross recommended an immediate operation through a vertical abdominal incision. He reduced the viscera into the abdomen, unrolled the edges of the defect, and closed the opening with interrupted silk sutures. He emphasized the association of intestinal malrotation and the small abdominal cavity. Dr. Gross closed only the skin to prevent increased intra-abdominal pressure and did not drain the pleural cavity [13].

From mid-century on, infants who developed symptoms after 24 h survived, but those diagnosed at birth continued to have a high mortality. Was this due to our failure to follow the advice of Dr. Gross on abdominal wall closure? Newborn infants rely on diaphragmatic respiration, and even minimally increased pressure will compromise gas exchange. Pressure on the vena cava reduces cardiac output. Many surgeons thought Dr. Gross's failure to mention a chest tube was an oversight, but rapid removal of air will not expand the ipsilateral hypoplastic lung. The mediastinum will shift and over-expand the contralateral lung. Newborn puppies survive with normal blood gases after a pneumonectomy, but if air is removed from the pleural cavity, the mediastinum shifts, and the contralateral lung overexpands with a rise in alveolar carbon dioxide, hypoxemia, and death [14]. During the early 1960's, I operated on two infants, symptomatic at birth, who did well for 12 h, then suffered increased respiratory distress. I re-opened the abdomen and closed only the skin. Both infants survived, and I closed the ventral hernias at one year of age. Like most surgeons, I drained the pleural cavity with a water-seal until a patient developed a contralateral pneumothorax and died within hours after surgery.

During the early 1970's, there was increasing frustration with the high mortality in infants under 24 h of age. They dramatically improved after surgery, then a downhill spiral led to death. This curious phenomenon was termed, the 'honeymoon period'. The introduction of long term endotracheal intubation and mechanical ventilation did not reduce mortality. In fact, when combined with pleural drainage, its use increased damage to the contralateral lung. Changes in anesthetic management included the use of muscle relaxants and narcotics, such as Fentanyl, a powerful respiratory depressant that can cause chest wall rigidity [15]. Rather than restore spontaneous respiration, anesthesiologists tended to leave the endotracheal tube in place and support the infant with a ventilator.

Research on fetal lambs suggested the mortality was due to 'persistent fetal circulation' with high pulmonary vascular resistance and a right to left shunt through the ductus arteriosus and the foramen ovale. These and other studies led to monitoring pre and post-ductal arterial blood gases and the use of drugs to dilate pulmonary vasculature [16,17]. Survival rates in critically ill patients improved, but prolonged endotracheal intubation, muscular paralysis, and sedation with mechanical ventilation brought on barotrauma, muscular wasting, and prolonged respiratory insufficiency.

In 1977, John German and Robert Bartlett at the University of California Irvine, opened a bold new approach to the management of diaphragmatic hernia with Extra-Corporeal Membrane Oxygenation, [ECMO]. Their four patients had low APGAR scores at birth and underwent immediate surgery. One patient had a contralateral pneumothorax following the operation, suggesting either over-zealous ventilation or mediastinal shift due to pleural drainage. Their patients showed transient postoperative improvement, then a downhill course with persistent fetal circulation, treated with vasodilator drugs, ligation of the patent ductus, and ECMO in three patients. One patient survived after seven and a half days on bypass [18].

Prolonged life support with extracorporeal oxygenation evolved from an effort by a German physiologist in 1885 to perfuse isolated organs by exposing a thin film of blood to oxygen in a rotating glass cylinder [19]. Charles Lindbergh, the aviator, with Alexis Carrel at the Rockefeller Institute during the 1930's, developed a successful apparatus for prolonged organ perfusion. They hoped to develop a means to support the heart