

# The triad of bridging bronchus malformation associated with left pulmonary artery sling and narrowing of the airway: the legacy of Wells and Landing

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**Abstract** Although noted in the 19th century, it was not until 1938 that Scheid published the combination of left pulmonary artery sling and narrowing of the airway due to annular tracheal cartilages. Unaware of these prior descriptions, and without a precise preoperative diagnosis, Willis Potts in Chicago performed the first successful sling repair in 1953. In 1976, Cohen and Landing described Scheid's combination of left pulmonary artery sling and stenosis caused by complete tracheal rings, and the term "ring-sling complex" was introduced by Berdon in 1984. Four years later, Wells and Landing noted characteristic tracheobronchial malformations associated with these lesions and proposed a classification that has been confirmed to be clinically relevant in recent cross-sectional imaging studies.

**Keywords** Bridging bronchus · Pulmonary artery sling · Tracheal narrowing · Wells · Landing

## Introduction

Pulmonary slings present as an airway disease in infants with stridor. During the last half century, it became obvious that this entity comprises a combination of different malformations of the pulmonary arteries, the airway configuration and the tracheal cartilage rings. About one-third of slings are characterized by an otherwise normal airway with a carina at the level of T4-5 and the left pulmonary artery wrapping around the trachea posteriorly above the right mainstem bronchus. However, two-thirds typically have a low-lying carina at T6-7 with airway narrowing and much of the right lung supplied by a bridging bronchus from the left bronchus, which the left pulmonary artery encircles posteriorly and back toward the left (Fig. 1); this latter group may cause significant morbidity and mortality in affected infants, and many may require repair of the stenotic parts of the airway. The airway narrowing may be due to compression of the anomalous artery, complete tracheal rings with lack of a membranous portion, malacia as a result of in utero compression of the developing airways or a combination of the above.

Although simple sling was described by Glaevecke and Doehle in 1897, the detailed description by German pathologist Paul Scheid in 1938, covers the first successful repair of a pulmonary sling by Potts, and moves on toward a discussion during the last half century on the complex classification introduced by Wells and Landing, and Zhong, who based her assessment on CT reconstructions.

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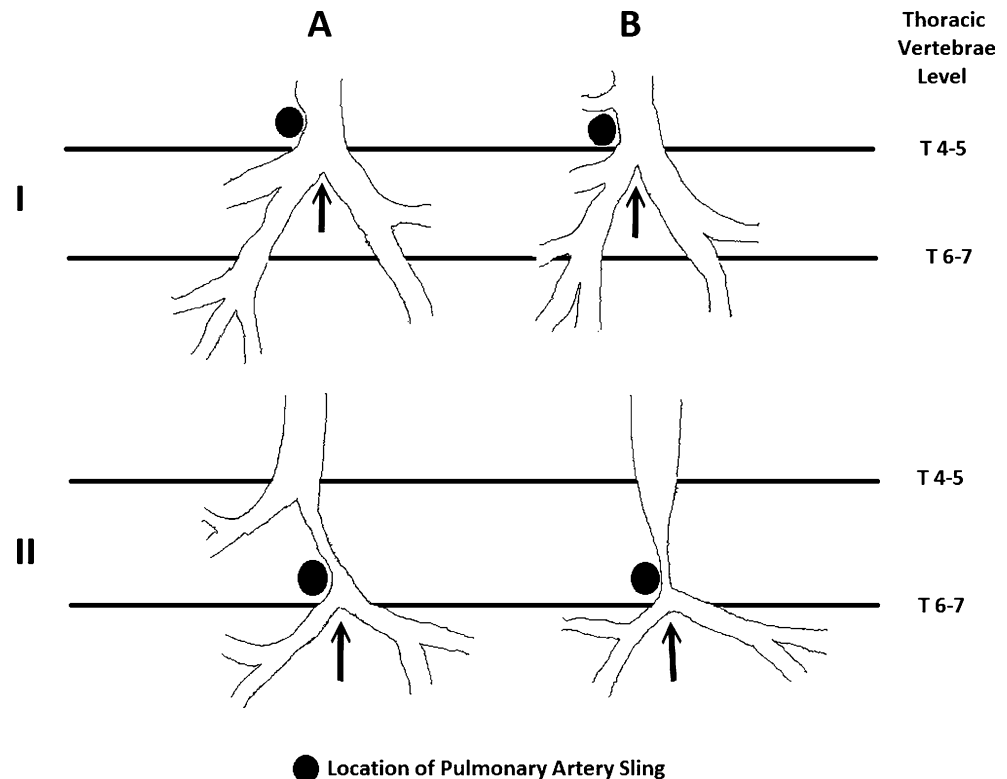
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**Fig. 1** Classification of airway malformations associated with pulmonary artery sling. Narrowing of the airway in type I malformations occurs by external compression of the pulmonary artery sling on an otherwise normal trachea. In type II malformations, intrinsic airway narrowing is present as a result of tracheal rings. Type IA has a normal configuration. In type IB, the right upper lobe is supplied by a right tracheal bronchus. The right lower lobe is supplied by a bridging bronchus in type IIA. The right upper lobe bronchus is absent in type IIB (adapted from [12] and [15]). The trachea is located at the level of T4-5 in type I and T6-7 in type II lesions. Carina = arrows



### Timeline

In 1897, Glaevecke and Doehle presented the case of an infant with congenital worsening stridor who died at age 7 months from a concomitant upper respiratory tract infection at the meeting of the Physiologischer Verein (Physiologic association) in Kiel, Germany. A short report from this presentation was published in *Münchener Medizinische Wochenschrift* [1]. The postmortem exam describes the findings of a left pulmonary artery sling with narrowing of the trachea, but it does not mention ring cartilages.

More than 40 years later, Scheid, a young pathologist-in-training at the University of Frankfurt in Germany, published the case of another 7-month-old boy who succumbed to worsening stridor and respiratory failure, providing a detailed description and sketch of the malformation, including the tracheal narrowing due to cartilaginous rings (Fig. 2) [2]. Of note, Scheid did not quote Glaevecke.

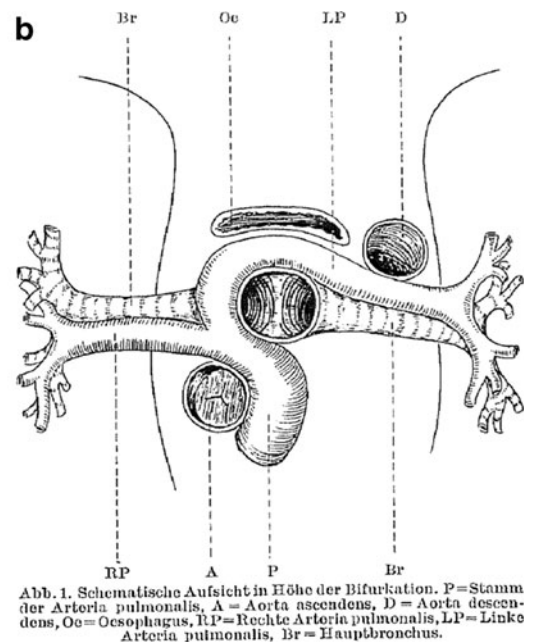
After World War II, Holinger, a member of an Ear, Nose and Throat surgical group in Chicago noted several young patients with stridor and extrinsic compression of the trachea on bronchoscopy. One of the children underwent thoracotomy by Willis Potts (Fig. 3) in 1953, who, without knowing of Glaevecke or Scheid, noted an anomalous left pulmonary artery coursing around the trachea during the operation. He transected the vessel and reanastomosed it anterior to the trachea, which alleviated the symptoms of

the child [3]. The case was republished in 1958 [4], at which time Scheid was referenced for the first time. Similar findings were reported in Boston at the same time by Wittenborg [5], who also noted the indentation of the esophagus on barium swallow and found that most of these patients had normal plain film chest radiographs.

During the 1960s, most published cases discussed simple and complex slings with and without intrinsic airway narrowing [6, 7]. In 1971, however, in a paper on pulmonary artery sling, Capitano et al. [8] published a complex case with a low carina in which the right lower lobe of the lung was supplied by a large aberrant branch coming off the left mainstem bronchus. This observation was later confirmed in another case and the name bridging bronchus was coined by Gonzalez-Crussi and collaborators [9] in Indianapolis.

The pathologist Benjamin Landing and the otorhinolaryngologist Cohen at the Children's Hospital Los Angeles found the combination of this bizarre tracheobronchial anomaly combined with left pulmonary artery sling and airway narrowing in children with rigid, unpassable airways during tracheobronchoscopy (Fig. 4) [10]. During the following two decades, Landing and his associate Theadis Wells, a laboratory assistant who later was appointed honorary clinical associate professor, noted the heterogeneity of these malformations (Fig. 4). Aware of the 1986 publication by Berdon et al. [11] on the ring-sling complex in a description of 5 patients with long-segmental tracheal stenosis due to cartilaginous rings and a low carina, Wells

**Fig. 2** **a** Paul Scheid was born 1907 in Koblenz. He studied medicine in Frankfurt and Berlin and trained in pathology for 3 years at the Westend hospital in Berlin, after which he moved to Frankfurt am Main where he completed his doctoral dissertation in 1938. He was drafted for military service from 1942 until 1944, and became chief of pathology at the Hospital Dresden-Friedrichstadt in 1949, where he worked until his retirement in 1974. He died of cancer in 1991 (Courtesy of Professor Gunter Haroske, Institute for Pathology, Krankenhaus Dresden-Friedrichstadt). **b** Illustration from Scheid's 1938 case of left pulmonary artery sling with tracheal stenosis [2]



and Landing proposed a classification in 1988 [12]. They also performed 3-D wax plate reconstructions of the airways based on CT images that show the profound airway narrowing associated with low carina (type II) anomalies [13].

Recent studies from Stanford University [14] and Shanghai, China, [15] using high-resolution contrast CT have corroborated Wells and Landing's classification,

defined the relationship of the airway and vascular anomalies, and shown that the numeric distribution among the different types is about equal.

**Discussion**

When Willis Potts performed the first repair of a pulmonary artery sling, he did not know about the previous descriptions by Glaevecke and Scheid. He essentially stumbled upon a relatively simple type IA malformation, and relieved the extrinsic compression of the trachea by transposing the left pulmonary artery anterior to the trachea.

In the following decades, historic pre-World War II reports resurfaced and were eventually integrated with newer descriptions on combined malformations of the airways and the left pulmonary artery into the landmark classification proposed by Wells and Landing in 1988. It became apparent that type II slings were a particular cause of morbidity and mortality, and that these could not be treated by reanastomosis of the pulmonary artery alone, but that the narrow airways most often required repair of the stenotic airway by tracheoplasty at the same time.

The technique of repair has evolved toward using cardiopulmonary bypass in most cases and a median sternotomy. The tracheal repair is accomplished by either pericardial patch tracheoplasty, tracheal autograft or slide tracheoplasty. In patients with absent or severely hypoplastic right lung, the left pulmonary artery is translocated anterior to the trachea. In the other patients, the left pulmonary artery is generally



**Fig. 3** Willis Potts was chief of surgery at Children's Memorial Hospital in Chicago. In 1953, he performed the first surgical correction of a pulmonary sling by reanastomosis anterior to the trachea in a 5-month-old boy after performing an exploratory thoracotomy for stridor and a narrow airway on tracheobronchoscopy. The stridor resolved, but on a follow-up study of the patient 24 years later, the left lung had no pulmonary arterial blood flow [16] (Courtesy of Dr. Carl Backer, Children's Memorial Hospital)



**Fig. 4** Photographs of Landing and Wells. **a** Dr. Benjamin Landing (1920–2000) was chief of pathology of Children’s Hospital Los Angeles for more than four decades. He had a particular interest in cardiac and thoracic anomalies. He was one of the acknowledged premiere pediatric pathologists in the world (Courtesy of Dr. Marvin Nelson). **b** The remarkable career of Theadis Wells (\*1938) started in Oklahoma City, where he graduated from Booker T. Washington High

School in 1956. He joined the U.S. Navy as a medical corpsman from 1956 to 1961. After working as an orderly at Mount Sinai Hospital in Los Angeles in 1963 and 1964, he took a job at Children’s Hospital Los Angeles in the histology laboratory, where he met Landing. His extraordinary abilities as a senior laboratory technician eventually earned him the appointment to clinical associate professor at the University of Southern California (Courtesy of Dr. Bruce Beckwith)

reimplanted into the main pulmonary artery. Using these techniques, the long-term patency rates of the pulmonary artery are excellent.

Despite the availability of cardiopulmonary bypass, the more complex type II associations therefore pose a considerable challenge in diagnosis and treatment. They require a multidisciplinary approach for preoperative planning and successful repair.

The imaging workup has evolved from bronchograms and angiograms to CT with intravenous contrast, which shows both the vascular anatomy and the airway configuration in fine details. In particular, coronal and sagittal reformatted images can be useful for preoperative planning, along with volume-rendered airway images. An esophogram is still a useful initial study for screening purposes and may show a typical indentation at the level of the sling. MRI has the drawback of motion artifact and lower resolution compared to CT. Also, the longer examination times limit the utility of MRI in unstable patients with airway compromise. The major drawback of CT is the relatively high radiation dose absorbed during the exam, although newer imaging protocols using 80 or 100 kVp afford good imaging quality despite lower radiation doses. For screening, it is unfortunate that techniques such as barium swallow and airway imaging, which

uses low-dose, high-kV, heavily filtered plain film techniques thereby decreasing radiation exposure in many children, are not commonly used anymore. It is important to note that plain film chest radiographs in most cases are either normal or in rare cases may show a variable degree of obstruction and pulmonary overinflation, usually right-sided.

Echocardiography can only visualize the vascular sling; it does not show the configuration or stenosis of the airways.

## Conclusion

The two entities of isolated pulmonary sling and high carina (types Ia, b) and pulmonary sling with low carina, airway narrowing and cartilage rings (types IIa, b) have been confounded for almost a century until a systematic classification was published by Wells and Landing in 1988. Modern spiral CT with intravenous contrast and coronal reconstruction is becoming the standard for the diagnosis of these complex anomalies.

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