Congenital Esophageal Atresia with Tracheoesophageal Fistula

A 4-lb, 5-oz premature female infant was born of a pregnancy complicated by maternal hydramnios. At birth and shortly thereafter, the infant showed an excessive amount of oropharyngeal secretion. Initial feeding twelve hours after birth resulted in vomiting, choking, and cyanosis. The abdomen was distended and became larger upon coughing and crying. The infant was subsequently transferred to an intensive care unit.

EXAMINATION

The possibility of esophageal atresia accompanied by tracheoesophageal fistula resulted in the introduction of a radiopaque catheter into the esophagus. The catheter stopped 9 cm from the upper gum margin. A roentgenogram was made to determine the level of the obstruction in relation to the vertebrae and to enable estimation of the length of the blind upper esophageal pouch. Due to the distended abdomen, a full-length air contrast roentgenogram was made, which indicated air in the stomach and intestines, thus establishing the presence of a fistulous communication between the trachea and the lower esophagus. The roentgenogram of the abdomen revealed no additional malformations and the infant was considered a good risk for a one-stage surgical repair.

DIAGNOSIS

Congenital esophageal atresia with tracheoesophageal fistula (Fig. 18C)
Tracheoesophageal fistula, arrow Trachea, T; upper esophageal pouch, U; ophageal pouch, LE.
TREATMENT

Following preoperative management and medication, the
child was placed in a lateral decubitus position with the right side up. The
uppermost Using an extrapleural surgical approach, the
thoracic wall was entered by the fourth rib and reflecting the parietal
vagus nerve, upper and lower esophageal pouches, and the fistula
connection between the lower esophagus and trachea were identified.
The lower esophageal pouch and the trachea were separated, and the
stability of the suture line on the trachea was achieved by
submerging the latter in a saline solution and applying pressure. The upper esophageal pouch was mobilized first by the lower esophageal pouch. Any gap between the two
lesions was overcome by additional mobilization of the pouch due to its greater vascularity and thicker wall. The upper
lesion of the lower esophagus was excised to produce a larger
creased blood supply, and a separate mucosal and muscularis
anastomosis was made to prevent recurrence of a fistula. There was well tolerated and postoperative feedings of glucose and
water were continued for three days. On the fourth day,
contrast swallow was performed to determine any leakage of
mucus and the passage of material into the stomach. With no
leakage, oral feeding of glucose and boiled skim milk was
begin on the fourteenth postoperative day. Bottle feeding began the third week. Except for several dilated
anastomotic sites, the patient made an uneventful recovery and returned home two weeks after admission.

DISCUSSION

The first known description of congenital esophageal at
traction of esophageal fistula was given by Thomas Gibson in 1897. Other cases, were not noted until the 1800's and, until the
teenth century, interest was concentrated on the embryology
with only sporadic attempts at treatment. In 1929, von
sent a classification, still in use, of this defect which affect
sely 1 of 2,500 to 3,000 newborn infants. Lanman was the first to attempt a primary anastomosis of the two
e segments and in 1943 Haight and Towsley were the first to
repair a primary extrapleural esophageal anastomosis. A mul
t repair was initiated by Ladd and Leven and in 1962 Ho
Donald, and Wooley\textsuperscript{17} reported increased success for critically ill and premature infants using a staged repair.

**Embryology**

Explain the occurrence of this malformation embryologically. Differentiation of the esophagus and the trachea occurs between the third and sixth week of gestation, at which time this malformation originates. In the 3-week-old embryo, a groove, the tracheobronchial sulcus, develops on the floor of the foregut between the thyroid anlage cranially and the liver primordium caudally. The epithelium of the foregut is one to two cells thick, whereas in the region of the tracheobronchial sulcus it is three or more cells thick. The ventral wall of the sulcus thickens as a result of epithelial proliferation and lateral ridges form in a coronal plane. These ridges, upon eventual fusion, separate the caudal portion of the developing trachea from the esophagus. This fusion gradually extends cranially. As a result of incongruence in growth between epithelial and mesodermal components, the epithelial cells are abundant and tend to fill the esophageal lumen. Later, as the mesoderm increases, the epithelium arranges itself along the wall of the esophagus and the width of the lumen increases. It is probable that this epithelial proliferation occurs throughout the digestive tract, especially in the esophagus, duodenum, colon, and rectum.

The various theories proposed for the occurrence of esophageal atresia and tracheoesophageal fistula include: (1) inflammation and ulceration of the developing esophagus and trachea; (2) deficiency of material for the complete construction of two separate systems; (3) relative pressure changes; (4) malposition of the large vessels, especially the right subclavian artery; (5) vascular insufficiency to the area; and (6) deviation of the normal course of the tracheobronchial sulcus, which begins at the site of tracheal bifurcation and eventually separates the trachea from the esophagus.

**Classification**

Classify the various forms of congenital atresia of the esophagus (Fig. 18). Numerous classifications exist; the best known is that of Vogt\textsuperscript{42}

- **Type 1.** Congenital agenesis of the esophagus.
- **Type 2.** Atresia of the esophagus without fistula.
- **Type 3.** a. Atresia of the esophagus with tracheoesophageal fistula of the proximal esophagus.
Fig. 18 Various forms of congenital atresia of the esophagus. Approximate given in percentage. (From Rickham, P. P., and Johnston, J. H.: Neonat Butterworths and Co., London, 1969, with permission.)
geal fistula of the distal esophagus.

c. Atresia of the esophagus with tracheoesophageal fistula of both the proximal and the distal esophagus.

The Vogt classification was modified by Ladd and by Roberts into that currently used:

1. Atresia of the esophagus without fistula.
2. Atresia of the esophagus with fistula between the proximal segment of the esophagus and the trachea.
3. Atresia of the esophagus with fistula between the distal segment of the esophagus and the trachea
   a. Proximal and distal esophageal segments are widely separated.
   b. Proximal and distal esophageal segments are close together or overlapping.
4. No atresia of the esophagus "H"-type tracheoesophageal fistula
5. Stenosis of the esophagus.

Surgical Approach

What are the advantages and disadvantages of the extrapleural and transpleural surgical approaches? The extrapleural approach presents less trauma to the lung, which remains behind the parietal pleura. If esophageal anastomotic leakage occurs, the infection remains localized and can be readily drained. The main disadvantages are the long operating time, limited exposure, frequent injury to the parietal pleura, and the reduced lateral motion postoperatively due to resection of a rib.

The transpleural approach allows a shorter operating time, wide field of exposure, and the use of mediastinal material for anastomotic suturing. The great disadvantage is that if an anastomotic leakage occurs, empyema and death result in a high percentage of the cases; also there is unavoidable trauma to the lung, which may produce atelectasis and subsequent pneumonitis.

Postoperative Complications

What factors may cause a complicated postoperative course? Pulmonary complications—i.e., atelectasis, pneumonia, pneumothorax, and empyema—anastomotic leakage, or stricture account for most postoperative complications.
What major malformations are frequently associated with atresia and trachoesophageal fistula? Atresia of other portions of the gastrointestinal tract, imperforate anus, lesions of the aorta, and polycystic kidney are closely correlated with this congenital
Agenesis of the Lung

A male infant, 8 months old, was admitted to Children's Hospital for wheezing and difficulty in breathing. Since birth these symptoms had gradually increased, but there was no cyanosis and prior to hospital admission he was treated for pneumonia. Physical examination showed the chest to be asymmetric. The left side was smaller and demonstrated diminished movement, dullness on percussion, and absence of breath sounds.

X-ray and fluoroscopy showed the entire left lung field to be opaque, while the right lung showed marked overexpansion. The mediastinum was shifted to the left side, which revealed narrower intercostal spaces than on the right. Thirteen ribs were present on each side.

A bronchogram showed no evidence of a left bronchus or its vestige.

The infant was bronchosoped under local anesthesia. The larynx was rotated toward the left, but the vocal folds moved normally. The trachea was displaced to the left; the left primary bronchus was totally obstructed about 1 cm. below the carina, whereas the right bronchial tree was normal. No other malformations were noted.

DIAGNOSIS

Agenesis of the left lung.

FURTHER COURSE

In the absence of other malformations, unilateral agenesis is compatible with normal existence, but the prevention or care of respira-
Above, X-ray picture showing agenesis of the left lung; Below, Bronchogram of a neonate showing little evidence of a left bronchus.
care for periodic checkups. Over the next four years he showed good development and his breathing difficulties subsided.

**DISCUSSION**

Complete agenesis of pulmonary tissue is extremely rare among accephalic monsters. Unilateral agenesis is not uncommon; more often in males, and the left lung is affected twice as often as the right. Morgagni described agenesis of the lung in 1762, and Munchmeyer was the first to attempt diagnosis of unilateral agenesis, later verified at necropsy. Using clinical evidence, Stokes and Brown (1940) made the first correct diagnosis. Bonnier (1928) based on bronchography, and Stokes and Brown (1940) made the diagnosis bronchoscopically.

**Embryology**

Discuss normal lung development. At three and one-half weeks' gestation (Fig. 19A), the respiratory system (larynx, trach...
growth from the floor of the primitive foregut. The grooves present fused pharyngeal pouches. The mesenchymal constituents of the respiratory system arise from splanchnic mesoderm on the ventral surface of the foregut into which the respiratory diverticulum and its subsequent subdivisions extend.

At four weeks (Fig. 19B), the caudal end of the tracheobronchial groove grows ventral to the foregut, forming an independent tracheal tube that is dilated at its distal end to form the primary bronchial buds. Soon the bronchial buds become asymmetrical, the left one lying more transversely than the right. Early branching of the primary bronchi tends to be monopodial (Fig. 20); i.e., a branch is formed on one side of the bronchi while the main stem continues to grow beyond this point without essential change of direction. Branching after the pri-

Fig. 20 Bronchial tree development: A, 28 days; B, 30 days; C, 32 days; D, 33 days; E, 35 days

mary bronchi are established is more likely to be dichotomous, i.e., bifurcation into two symmetrically placed branches, neither of which extends in the direction of original growth. At this stage (Fig. 20E), the right lung bud possesses three bronchial tubes, while the left has two. These endodermal tubes, together with the lung bud mesenchyme that surrounds them, give origin to the lobes of the adult lungs. The part of the foregut dorsal to the trachea forms the esophagus and is
ends of the esophagus are fixed in position.

Once the primary bronchi are established, four stages of tree development occur:

1. **Glandular**—to the fourth month of gestation, during which the remaining divisions of the bronchial tree are formed, respiratory-like movements ensue.

2. **Canalicular**—from the fourth to the sixth month, the tubular portion is delineated. The lung mesenchyme becomes vascular and capillary loops surround the developing tubules.

3. **Alveolar**—from six months to term, branching continues, generations of alveoli, which is the characteristic number of adult lung, are formed. By the seventh month, capillaries surrounding the alveoli are sufficiently developed to sustain intrauterine existence. As additional alveolar sacs form, a barrier separating the blood of the pulmonary circuit from the alveolar air becomes sufficiently thin to readily permit of O₂ and CO₂.

4. **Birth**—the terminal bronchioles continue to divide postnatally; soon after birth, six or seven additional generations; this occurs before the first eight years of life. At birth, the initiation of the bronchiole dilates and expands the alveoli. This expansion begins with the vascular central region of the lung and at approximately three days of age, the alveoli of the peripheral areas are dilated.

A lipoprotein compound, which coats the epithelial surface of the alveoli and contributes to the stabilization of the alveolar air, is of crucial importance.

Fetal aspiration of amniotic fluid into the lungs may occur, thought to result only from asphyxia or a similar condition, and normally present in the fetal alveoli and tracheobronchial tree. Aspirated amniotic fluid, but an ultrafiltrate of plasma.

**Causes of Lung Agenesis**

Several factors are proposed as the cause of lung agenesis, include disturbances in: (1) genetics; (2) fetal nutrition; and (3) opamental mechanics. Additional causes may include interference with the fetal blood supply and disturbances in closure of pleural communications.
What lung malformations or disease can interfere with the diagnosis of lung agenesis? Atelectasis, bronchiectasis, postpneumonectomy chest, fibrothorax, and blockage of a main bronchus by tumor give symptoms similar to those of agenesis. These may be eliminated using bronchoscopy and bronchogram.

Classification

Two sets of classifications have been used to describe lung malformations:

1. Schneider’s Classification\(^\text{27}\)
   a. Agenesis—complete absence of a lung, bronchus, or vascular supply
   b. Aplasia—presence of bronchial rudiments without alveolar tissue
   c. Hypoplasia—deficient growth of regularly differentiated pulmonary components

2. Minetto’s Classification\(^\text{27}\)
   a. Bilateral pulmonary agenesis
   b. Unilateral pulmonary agenesis
   c. Pulmonary aplasia
   d. Pulmonary hypoplasia
   e. Lobar aplasia

Associated Malformations

What malformations are most commonly associated with lung agenesis? Narrowed trachea, extracartilaginous rings, supernumerary bronchi of the normal lung, absence of pleura, tracheoesophageal fistula, and esophageal stenosis are most frequently found with lung agenesis. In addition, defects of almost every tissue and organ have been reported in conjunction with agenesis.
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Patent Ductus Arteriosus

An 18-month-old acyanotic female is admitted to Children's Hospital for cardiac catheterization. Her physical development (height and weight) is retarded; mental development is normal. Recently, a heart murmur was detected during treatment for pneumonia. Upon admission, a continuous characteristic (“machinery”) I murmur, accentuated during systole, is heard in the second intercostal space to the left of the sternum. A thrill is present over the precordium. Radiologic examination reveals a slight left ventricular hypertrophy with an increase of lung vascularity. Cardiac catheterization performed on the fifth day following admission gives direct measurements of blood flow and confirms the diagnosis of patent ductus arteriosus with a left-to-right shunt. Surgery is indicated.

Family history reveals that the mother had rubella in the first few months of pregnancy.

DIAGNOSIS

A medium-sized patent ductus arteriosus (PDA)

TREATMENT

With the infant under general anesthesia, a posterolateral incision was made in the fourth interspace and the mediastinal pleura separated between the phrenic and vagus nerves. The vagus was retracted to expose the recurrent laryngeal nerve where it passes behind the ductus. Dissection began close to the aorta, removing the pericardium back to the pulmonary artery. Potts clamps were applied to the two ends.
wall was closed, recovery was uneventful, and on the ter-
active day examination revealed the absence of a precordi-
murmur; roentgenologic evidence showed a decreasing vs.
heart shadow and of lung vascularity.

DISCUSSION

Although originally described by Galen\textsuperscript{10} as a short \textit{v}
the aorta and the pulmonary artery, the ductus arteriosus i
associated with the name of Botallo (seventeenth century
1766 von Haller\textsuperscript{10} translated the published works of Botallo
one of his own drawings depicting a connecting vessel
aorta and the pulmonary artery. Gibson (1900)\textsuperscript{11} describe
classic "machinery" murmur, and in 1907 Munro\textsuperscript{28} suggest
approach and operative closure of the patent ductus. Gray
and Boyer (1938)\textsuperscript{18} attempted closure of an infected p

![Diagram of heart and arteries]

*Fig. 21 Anatomical relationships of a ductus arteriosus in the newborn*
Definition

Ductus arteriosus: The ductus arteriosus, a thick-walled arterial channel connecting the left pulmonary artery with the aorta immediately distal to the origin of the left subclavian artery. Patent ductus arteriosus usually occurs in one individual out of every 3,330 in the general population. The ductus arteriosus is a channel through which blood bypasses the lungs during the fetal period and it closes spontaneously shortly after birth. The mechanism of closure is debated, but probably at the onset of respiration, a drop in pulmonary vascular resistance results in a pronounced constriction of the ductus. Closure of the ductus is complete by fragmentation of the internal elastic lamina, allowing the smooth muscle to contract into the lumen. Patency then converts the ductus arteriosus into the ligamentum arteriosum.
Discuss embryologically the development and subentiation of the aortic arch arteries (aortic arches) and explain the occurrence of the ductus arteriosus. The primitive arteries are the right and left primitive aorta. They are early somite embryos (Fig. 22) as two vessels that arise from the endocardial tubes. Each passes from the floor of the foregut around the pharynx and to the side of the gut as the dorsal aorta. Each primitive aorta is divided into three portions: (1) an ascending, (2) pharyngeal arch, and (3) descending. Fusion of the two endocardial tubes (Fig. 23A) reaches the limit of the pericardial cavity and extends cranially to include the extremities of the two ascending portions. The latter forms a single midline vessel, the truncus arteriosus (Fig. 23B), which becomes dilated (three and one-half weeks) and is known as the aortic sac. The right and left first aortic arches arise from the development of the aortic sac. Successive pairs of aortic arches develop in association with each forming pharyngeal arches as communication channels between the aortic sac and the body cavity. Shortly after the appearance of the aortic sac and the first arch, the dorsal aortae undergo two changes:

1. Each aorta grows in a cranial direction beyond which it is joined by the right and left internal carotid arteries.
2. The dorsal aortae fuse caudally to form a single aorta, which lies between the developing somites and the notochord dorsally. As the aorta matures, the fusion extends forward, but not to the pharynx.

By the end of the first month of development (Fig. 2), the first arch disappears as the third arch fully differentiates. The fourth arch involutes as the fourth arch matures. The distal part of the first arch becomes the maxillary artery, while the distal part of the second arch forms the stapedial and hyoid arches. The proximal portion of the first and second aortic arches form part of the external carotid artery, and the aortic arch from its origin at the aortic sac to the arch of the aorta.
sequent differences). Aorta, and embryonic identified in ear as con-
trasting the region with the aspect divided into:
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Fig. 23 Sagittal view of aortic arch development: A, 4-week-old embryo; B and C, enlargements of area outlined in A. B, 4-week-old embryo; C, 6-week-old embryo.
fifth aortic arch exists transiently. The sixth aortic arch into a ventral rudiment that forms the pulmonary artery, rudiment that arises from the dorsal aorta and joins the aort, as the ductus arteriosus.

By the sixth week (Fig 23C), primarily because of vascular hemodynamics, the arterial system completes and undergoes modifications to the adult pattern. The first and sixth aortic arches are developed, and the bulbus aortic arch longitudinally divided into two parts by the growth of bulbar septum. The distal part of the septum extends into the sac such that blood which enters the bulbus from the right empties into the dorsal portion of the aortic sac and is aortic arch only; blood from the left ventricle empties into the cranial portion of the aortic sac and is distributed to the third aortic arches. Disappearance of the cranial portion of the aorta between its attachment to the left dorsal aorta and fourth aortic arch subsequently results in the loss of the do
is divided by a dorsal entral rudiment.

changes in formation of the sixth and fourth ventricle of the sixth and fourth right dorsal.

of the right sixth aortic arch beyond the origin of the pulmonary artery. This dorsal rudiment corresponds to the ductus arteriosus of the left side. The fourth aortic arch persists bilaterally; on the left it joins with the aortic sac and the left horn of the aortic sac to form the arch of the aorta up to the point of attachment of the seventh segmental artery, the latter which forming the left subclavian artery. On the right, the horn of the aortic sac becomes the brachiocephalic artery, which forms the stem of the right common carotid and subclavian arteries; the remainder of the subclavian is formed by the right fourth aortic arch (Fig. 24)

Fig. 24 Comparison of aortic arch system (frontal view): A, 1st arch artery (maxillary); B, External carotid; C, 2nd arch artery (stapedial, hyoid); D, 3rd arch artery; E, Common carotid (3rd arch); F, First part internal carotid artery (3rd arch); G, Ductus caroticus (dorsal aorta between 3rd and 4th arch); H, Brachiocephalic (aortic sac); I, 4th arch artery; J, 4th arch at attachment of 7th intersegmental (subclavian); K, Ductus arteriosus (dorsal rudiment of left 6th arch); L, Right subclavian; M, Transient (I) 5th arch artery; N, Ventral rudiment 6th arch artery (left pulmonary); O, Pulmonary trunk; P, Dorsal rudiment of 6th arch on right; Q, Right pulmonary artery; R, Head segment of internal carotid; S, Fused dorsal aorta; T, Aortic sac; U, IX nerve; V, Superior laryngeal n (X); V', (X); V', Recurrent laryngeal (X); W, Vertebral (longitudinal anastomosis of dorsolateral intersegmentals); X, Thyrocervical trunk; Y, Costocervical trunk; Z, Internal thoracic.
What heart or arterial malformations may be confused with patent ductus arteriosus? A PDA is most frequently confused with aorticopulmonary window (congenital communication between the ascending aorta and the stem of the pulmonary artery due to semilunar valves, resembling hemodynamically and provides a PDA). Other conditions that must be distinguished are pulmonary arteriovenous fistulas, ventricular septal defect, sufficient, ruptured sinus of Valsalva, and coronary arterial fistula.

Associated Malformations

What malformations are frequently associated with aortic stenosis of the aorta and ventricular septal defect are frequently associated malformations. In conditions such as pulmonary stenosis of Fallot, and pulmonary or tricuspid atresia, PD helps in keeping the child alive.