A 2-year-old boy was admitted to the hospital for evaluation of a heart murmur previously detected at birth. He was less active than other children his age, but, although overexertion was followed by frequent squatting and cyanosis of the lips and nails, there was no history of loss of consciousness. Initial examination revealed a thin, physically retarded, cyanotic child with no respiratory difficulty. Moderate clubbing of fingers and cyanosis of the nail beds was noted. A harsh systolic murmur was maximal over the mid-precordial area; no thrill was felt. The first heart sound was normal while the second was single, distinct, and loud. The lungs were clear. Roentgenologic study showed a normal-sized heart dominated by a boot-shaped (cœur en sabot) right ventricular outflow-tract. Peripheral vascular shadows in the lungs were less prominent than normal. Electrocardiography indicated a right axis deviation of the heart, while angiocardiography showed a right-to-left shunt through a ventricular septal defect, early appearance of dye in the ascending aorta due to its overriding position, and diminished blood flow through the lung fields.

**DIAGNOSIS**

Tetralogy of Fallot by definition consists of a ventricular septal defect and pulmonic stenosis (valvular or infundibular) resulting in an overriding aorta and right ventricular hypertrophy (Fig 25)
Fig 25 Blalock-Taussig shunt

TREATMENT

When the child was 2 years of age, a palliative Bla shunt (Fig. 25) was performed by anastomosis of the divid end of the left subclavian artery to the side of the left artery, resulting in increased collateral circulation to the hu relieving the cyanosis. Open intracardiac repair was perfo the child was seven years old, with closure of the palliati channel, relief of the pulmonary outflow-tract obstruction plete closure of the ventricular septal defect. The patient complicated recovery and is acyanotic and asymptomatic on-

DISCUSSION

Six out of 1,000 live births are affected by anomalies of t great vessels. Stensen in 1671 first described the ar was to become known as tetralogy of Fallot. In 1888 Fa
the four basic components comprising the tetralogy. Diagnosis and palliative treatment advanced under Blalock and Taussig (1945) and under Potts, Smith, and Gibson (1946). In 1955 Lillehei and coworkers and Kirklin and colleagues began total open intracardiac correction of tetralogy of Fallot utilizing a cardiopulmonary bypass.

Embryology

Discuss developmentally the defects that characterize tetralogy of Fallot. In the late presomite embryo, the angiogenic tissue from which the primordium of the heart will develop first appears as scattered groups of cells arising in the mesenchyme of the anterior margin of the embryonic disk cranial to the neural plate; this is the cardiogenic area. These groups of angiogenic cells soon fuse into a plexus of endothelial vessels which unite to form right and left endocardial heart tubes; the latter gradually merge to establish a single median endocardial tube. At four weeks of gestation, the primitive heart is not divided into separate chambers, but undergoes differential expansion that results in several dilatations separated by grooves. From caudal to cranial, the dilatations are the sinus venosus, the atrium, the ventricle, and the truncus arteriosus. Subsequently, a major change occurs with the partitioning of the heart tube by the formation of the cardiac septa.

Early in the second month (Fig. 26A), indication of the division of the primitive ventricle into right and left chambers occurs with the development of the primary muscular partition of the interventricular septum. This crescent-shaped septum appears at the apex of the ventricular bend and is directed toward the anteroventricular canal cushions (dorsal and ventral endocardial cushions which separate the atia from the ventricles). An interventricular foramen persists for a short time between the interventricular septum and the canal cushions; this foramen is closed by material derived from several sources (i.e., conus ridges, endocardial cushions, and the crest of the original interventricular septum).

Truncus ridges between the roots of the fourth and sixth aortic arches pursue a spiral course through the truncus arteriosus toward the ventricles, and form a complete partition separating an aortic channel leading into the fourth aortic arches from a pulmonary channel leading into the sixth arches (Fig. 26B). The ascending aorta and the main pulmonary trunk (Fig. 26B) twist around each other as they emerge from the ventricles, thereby bringing the aortic and pulmonary channels into contact with the left and right ventricles, respectively. The level at which the aortic and pulmonary valves develop is regarded as the
Fig. 26 Internal development of the heart. A, 5-week-old embryo; B, 3-

line of demarcation between the truncus arteriosus and ventricular outlet; this is the conus arteriosus (infundibul) ventricular side of the aortic and pulmonary valves, the c the truncus ridges form the conus ridges. The conus rid; the crest of the interventricular septum, thereby reducing the interventricular foramen. Local enlargements of the cushions and of the connective tissue that caps the crest of portion of the interventricular septum complete the closure of ventricular foramen. At the site of final closure, the interventricular septum gradually becomes a thin fibrous sheet called the trabecular portion (Fig. 25E). The tetralogy of Fallot results from motion of the distal portion of the pulmonary conus ridge at
with inadequate posterior-septal growth of the muscular portion of the interventricular septum and incomplete formation of the membranous portion. Thus, the interventricular foramen is retained and accompanied by infundibular stenosis. With incomplete formation of the ventricular septum, the base of the aorta shifts to the right and overrides the muscular portion of the interventricular septum, allowing the aorta to communicate with both right and left ventricles. Infundibular stenosis results in right ventricular hypertrophy with a right-to-left shunt.

Palliative Procedures

What are the most common palliative procedures employed in children under 5 years of age? In the Blalock-Taussig shunt used in the
present case, the Potts procedure (Fig. 27), which is between the left pulmonary artery and the aorta, and this direct correction of the pulmonary obstruction are alternative procedures. The main disadvantage of certain is the difficulty with which they are dissected and closed.

total correction

Associated Malformations

The most frequently associated major malformations are absence of the pulmonary artery and complete situs malformations are an absence of the medial papillary of the tricuspid valve, and occurrence of a right-sided
Atrial Septal Defect

A 6-year-old girl first demonstrated a cardiac murmur at 4 years of age. Growth and development were slightly retarded with some limitation of exercise tolerance. Cyanosis and clubbing were absent. The frequency of upper respiratory infections increased between the ages of 4 and 6.

EXAMINATION

Physical examination revealed a slight asymmetry of the thorax with prominence of the precordium. The heart was not markedly enlarged and no thrills were palpable. A systolic murmur was audible along the left sternal border with maximal intensity in the second left intercostal space. The electrocardiogram showed a right axis deviation and incomplete right bundle branch block. Fluoroscopic examination disclosed: (1) dilatation of the right heart and pulmonary arteries and veins; (2) pulsation of the pulmonary artery branches; and (3) increased vascularity of the lung fields. Cardiac catheterization verified the diagnosis of atrial septal defect and contributed pertinent information regarding the size and localization of the defect.

DIAGNOSIS

Atrial septal defect—ostium secundum type.

TREATMENT

Employing direct vision and hypothermia, the chest was opened through a split-sternal incision. Tapes were placed on the superior and
interior venae cavae, and the vessels were closed with
quets and the heart emptied by massage. An surrounding
replaced by carbon dioxide during the cardiotomy. Th
was opened by a longitudinal incision and the interior inci
ing a 4 cm typical ostium secundum septal defect. VW
visualized a suture was placed in its superior margin to
the borders and to insure accurate placement of a suture
margin of the defect. The atrial septal defect then was
double continuous running suture that included the atr
atrium to seal it effectively from the opening of the infer
The caval tapes were removed and the atrial wall closed
stitches. The initial incision was closed by interrupted.
The patient was asymptomatic 3 months postoperative
catheterization revealed no left-to-right shunt at the atr

DISCUSSION

Atrial septal defects occur in 7 per cent of all cor
malformations. 29 Von Ecker (1839) 8 and Cruveilhier (18
dilatation and hypertrophy of the right heart, marked
pulmonary artery and its branches, and the relatively s
left ventricle were distinguishing features of the heart b
atrial communication. Nearly a century later, Cohn
artificially created atrial septal defects in dogs, while M
using a blind method of closure, was the first to opera
subject. Subsequently, several techniques of repair ha
duced. 1 7 80 41 Today the open-heart operation with d
most used, with some differences of opinion regarding
of cardiac bypass to utilize, the pump-oxygenator or hi

Embryology

Describe the partitioning of the primitive atrium.
primitive atrium becomes divided into right and left e
formation of two septa (Fig. 28E). Based on their se
ance, the septa are called septum primum and septum

In the second month of gestation (Fig. 28A), the s
a crescentic ridge on the dorsocephalic part of the at
ward the atioventricular canal. Concomitantly, two l
(endocardial cushions) appear in the anterior and po
the atioventricular canal. During the sixth week (Fig

114
The common nubers by the atial appear-
undum primum, a wall, grows
rion walls of B), the endo-
cardial cushions fuse, forming a common atrio-
cellular canal into right and left channels.

Between the concave margin of septum primum and the endocardial cushions is a progressively diminished opening known as the ostium primum (interatrial foramen primum). As septum primum fuses with the endocardial cushions, ostium primum is obliterated; at the same time a new opening appears in the septum primum—the ostium secundum (interatrial foramen secundum).

In the seventh week (Fig. 28C), septum secundum forms in the roof of the atrium and to the right of septum primum. Like septum primum, it is crescentic in shape, but never forms a complete partition in the atrium. Septum secundum grows downward following the base of septum primum, and its concave margin overlaps the ostium secundum, leaving an oval aperture known as the foramen ovale (Fig. 28D). The margin of septum secundum constitutes the adult limbus or annulus fossae ovalis.

Developmental arrest during septal formation accounts for the

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**Fig. 28 Partitioning of the atrium:** A, 5-week-old embryo; B, 6-week-old embryo; C, 7-week-old embryo; D, 3-month-old fetus; E, Relationships of septum primum and septum secundum during later fetal life.
Fig. 28 Continued. (See p. 115 for legend.)
Fig. 29 Classification of interatrial septal defects: A, Persistent ostium secundum; B, Persistent ostium primum; C, Patent foramen ovale.
variety of atrial septal defects. In general, interatrial
two-ventricle septum. Provided they do not include the atrioventricular part.
Interatrial septal defects make the individual more vul-
nerable to secondary disturbances.

Classification

In what way(s) may interatrial septal defects be
categorized by location (Fig 29):

1. Patent foramen ovale
   a. Uncomplicated
   b. Complicated
      i. Pulmonary stenosis
      ii. Anomalous pulmonary vein drainage
      iii. Mitral stenosis

2. Persistent ostium primum
   a. Central—often fenestrated
   b. Low—has no inferior margin

3. Persistent ostium secundum

4. Localized single or multiple defects anywhere
   septum

Differential Diagnosis

What malformations may give symptoms similar
defects? Atypical patent ductus arteriosus, pulmonary
malous pulmonary veins, and atrioventricularis commun
confused with atrial septal defects and are diagnosed a
from atrial septal defects using a variety of methods.

Treatment

What types of closure are employed for atrial septal
fixion of the atria using sutures in the plane of the sept
of the atrial appendages or of the lateral wall of an au
defect; opening the heart and closing the defect under di-
ing a pump-oxygenator to divert the blood; and tempo-
to 4 minutes) of the venae cavae and opening the at-
defect are methods of closure. In addition, atrial septa
paired using a rubber "well" attached to the right atr
directly through a pool of blood.

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Associated Malformations

What malformations most commonly accompany atrial septal defects? Partial anomalous pulmonary venous return, pulmonary stenosis, mitral valve stenosis, tricuspid atresia, and transposition of the great vessels are most frequently found.

REFERENCES


Congenital Diaphragmatic Hernia

A 22-hour-old male infant born at term (3,200 gm.) of an uncomplicated pregnancy was transferred to Children's Hospital because of respiratory difficulty (dyspnea).

EXAMINATION

Examination showed the infant to possess subcostal and sternal retractions, scaphoid abdomen with a lack of bowel sounds, and a slight cyanosis that was relieved by oxygen. X-rays revealed gas-filled intestinal loops in the left thorax accompanied by a mediastinal shift to the right, and hypoplasia of the left lung.

DIAGNOSIS

Congenital left posterolateral diaphragmatic hernia through the foramen of Bochdalek.

TREATMENT

Nasogastric suction was applied prior to surgery to help decompress the intestines. An abdominal incision revealed the small and large intestines up to the transverse colon displaced through a defect in the posterior left half of the diaphragm (Fig. 36). Exploration of the abdominal area indicated incomplete intestinal rotation. The abdominal viscera were subsequently removed from the left pleural cavity by gentle pulling and placed in an approximate normal position in the abdomen. No remnant of a hernial sac was noted. The diaphragmatic defect was repaired in two layers (pleural and peritoneal), and the abdominal wall
X-ray study of a left posterolateral diaphragmatic hernia showing gut loops in the left thorax, arrow.
Fig. 36 Above. Displacement of abdominal viscera in a left-sided diaphragmatic hernia; Below, Commonest diaphragmatic defects through which hernias occur
was closed with some difficulty. Postoperatively, expansion of the left lung did not occur and the patient expired.

DISCUSSION

Ambroise Paré (1575) gave the first description of a diaphragmatic hernia, which occurred in an adult. The pathology and classification of the various diaphragmatic hernias were described by Sir Astley Cooper (1844). In 1848 Bochdalek clarified the embryology of the type of diaphragmatic defect discussed in the present case. This defect is responsible for 1 in 50 neonatal deaths in the first three days of life and comprises 8 per cent of all major fatal congenital anomalies.

Embryology

Developmentally, how do you explain the present diaphragmatic defect? A hernia through the posterolateral foramen of Bochdalek involves three intimately timed embryological processes: (1) formation of the diaphragm that separates the pleural from the peritoneal cavity; (2) return of the midgut to the abdominal cavity from the umbilical stalk; and (3) lung formation. In the seventh week of development, the midgut undergoes a rapid growth; since the fetal abdomen is two small to contain the entire midgut, the latter passes into the umbilical stalk where it continues the processes of complex rotation around the superior mesenteric artery. By the tenth week the midgut returns to the left side of the abdomen and continues its rotation, with part of the midgut eventually coming to lie on the right side.

The following explanation of the formation of the intraembryonic coelom and the body cavities is essential to an understanding of the present case.

In an early somite embryo (Fig 37A), small spaces lined with mesothelium appear in the lateral plate mesoderm. These spaces prolongate cranially within the embryo, become confluent, and eventually fuse across the midline in the cardiogenic mesoderm. The resulting horseshoe-shaped closed cavity is the intraembryonic coelom. Later, at the lateral edges of the caudal extremities of the intraembryonic coelom, a communication with the extraembryonic coelom will ensue. The intraembryonic coelom is lined by splanchnopleure and somatopleure (Fig 37B).

The cranial portion of the intraembryonic coelom crosses the midline of the embryonic disk, transversely and anterior to the buccopharyngeal membrane, to form the pericardial cavity. The cardiogenic plate
develops in the splanchnopleuric mesoderm, and the tubes develop from this plate and designate the site of the cardiac cavity.

With the formation of the head fold, the endoderm becomes depressed into the embryonic disc to form the primitive gut. The splanchnopleuric mesoderm enters the gut and surrounds it. The somatopleuric mesoderm is then invaded by the gut and is displaced laterally. The intermediate mesoderm is formed by the folding of the splanchnopleuric mesoderm into the primitive gut and is the source of the kidneys and connective tissue of the alimentary tract.

Fig. 37 Division of coelom: A, X-s 3-week-old embryo; B, Dorsal view of 3-week embryo; C, X-s 5-week-old embryo; D, Sagittal view, 5-week embryo; E, Sagittal view, 6-week-old embryo; F, Sagittal view, 7-week-old embryo. (From Hamilton, W. J., Boyd, J. D., and Moss, A., Embryology, ed. 3. W. Heffer & Sons, Cambridge, England, 1946.)

(Illustrations are continued on the three following pages.)
endothelial heart
of the future peri-
chondrial heart tubes

AMNION
NEURAL GROOVE
SOMITE
INTRAEMBRYONIC
COELOM
EXTRAEMBRYONIC
COELOM
AMNION
NEURAL PLATE

Fig. 37 (continued. Illustrations continue on the two following pages. See facing page for legend.)
and the transverse pericardial portion of the coelom, both of which originally lay in the cranial margin of the embryonic disk, are bent ventrally and caudally beneath the foregut. Consequently, the original cranial edge of the embryonic disk is situated caudal to the pericardial cavity, and the original dorsal surface of this cavity faces ventrally. At this stage (fifth week) (Fig. 37C, D), the pericardial cavity is proportionately quite large and communicates freely with the peritoneal cavity via the pericardial-peritoneal canals. The mesoderm (fused splanchnopleure and somatopleure), which initially was cranial to the pericardial cavity, is now caudal to it and ventral to the developing foregut. This mass of mesoderm is the septum transversum. The pericardial cavity is bounded caudally by the septum transversum, in which the liver develops and through which the vitelline and umbilical veins pass to the sinus venosus.

Fig. 37 (continued Illustrations continue on facing page See page 146 for legend)
After formation of the head fold, the pericardial cavity remains in communication with the intraembryonic coelom by a constricted canal, the coelomic duct. This duct passes dorsal to the septum transversum. As the foregut lengthens, the coelomic ducts also elongate; each duct is invaginated on its medial aspect by a lung bud and the ducts become the pleural cavities. As the lung buds develop, the heart descends relative to the other structures, so that the common cardinal veins (duct of Cuvier) pass in a ventromedial direction to the sinus venosus (Fig. 37E), forming the pericardiopleural membrane that divides the pericardial from the pleural cavity.

On each side of the embryo, the peritoneal cavity freely communicates with the extraembryonic coelom. The early position of the gut and its mesentery in the midline divides the peritoneal cavity into a right and a left half. These halves intercommunicate ventral to the
gut (Fig 37D). With expansion of the peritoneal cavity and narrowing of the umbilicus, the communication between the intra- and extra-embryonic coelom is progressively constricted and finally obliterated as the umbilical cord loses its communication with the peritoneal cavity. Concurrently, as the pericardial cavity is closed off from the intra-embryonic coelom, the mesonephros becomes situated more caudally in the abdominal region. A ridge of mesoderm, the pleuroperitoneal membrane, connects the cranial pole of the mesonephros and the adrenal gland to the edge of the septum transversum. As the pleural cavity enlarges, the pleuroperitoneal membrane becomes extensive and eventually will close off the pleural from the peritoneal cavity (Fig. 37F).

After closure of the pleuroperitoneal communication at approximately 10 weeks, the resulting diaphragm (Fig 38) between the thoracic and abdominal cavities consists of:

1. An anterior central portion that represents the greater part of the original septum transversum; in the adult the septum transversum is the central tendon of the diaphragm.
2. Paired dorsal portions of the pleuroperitoneal membrane. The anterior margin of this membrane is continuous with the posterolateral edge of the central tendon.
3. A dorsal portion derived from the esophageal mesentery.
4. An extension of the lining and muscular tissue from the body wall.
Premature return of the midgut to the abdominal cavity or delayed closure of the diaphragm may produce a diaphragmatic hernia. The inter-relationship of the developing diaphragm and the gastrointestinal tract determines the type of diaphragmatic hernia and explains the frequent association of diaphragmatic hernias with malrotation of the gut. Since both bronchial and alveolar development of the lung lags behind development of the midgut, the severity of pulmonary hypoplasia (defective or incomplete development) is determined by whether the midgut passes into the thorax at the eighth (severe) or twelfth (mild) week of development.

Classification

A diaphragmatic hernia through the foramen of Bochdalek constitutes a surgical emergency in the newborn. What other diaphragmatic defects, congenital or acquired, are recognized? Hernias of the diaphragm have been categorized; the classifications differ slightly, but most investigators are in general agreement on the following types (Fig 36B): (1) retrosternal or retrocostosternal hernias through the foramen of Morgagni; (2) posterolateral defects; (3) eventration; (4) esophageal hiatus hernia; (5) extensive defects involving most or all of the hemidiaphragm; and (6) herniation into the pericardium.

Surgical Approach

Surgical approach to the hernia in the present case was abdominal. What are some advantages or disadvantages of an abdominal as compared to a thoracic approach? The abdominal procedure allows exploration for additional malformations of the gastrointestinal tract. Normally, the abdominal cavity needs to be stretched to accommodate the herniated intestine; this is best done utilizing an abdominal incision or by attachment of a Silastic sheet (nylon sheet covered on both sides with a layer of silicone rubber) to the edge of the abdominal incision if the herniated contents are too large to fit into the abdominal cavity. In neonates an abdominal approach is practical to determine whether the gut will fit into the abdomen; however, a surgical approach through the thorax immediately relieves pressure on the lungs, and thoracic malformations as well as a hernial sac, if present, are recognized. Closure of a thoracic incision is easy and secure, with fewer wound complications than with an abdominal approach. If necessary, a thoracic incision can be extended to the abdomen.
What malformations are most commonly associated with lateral diaphragmatic hernia? Abnormal fixation and hypoplasia of the lung on the involved side, with a diaphragmatic hernia. Malformations less gross defects of the neural axis, skull, and vertebra.