39. Pediatric Surgery

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General Considerations

The normal adaptive demands upon infants and young children are enormous. When compensatory requirements imposed by illness and operation are superimposed, the need for vigilant and meticulous care becomes apparent.

Fluid and Electrolyte Balance. In an infant or child the margin between dehydration and fluid overload is small. The infant is born with a surplus of body water, but within a few days this is excreted. At birth and for the first 10 days of life, fluid requirements are between 65 and 100 mL/kg (750 to 1.000 mL/mm^2). Maintenance fluids can be conveniently calculated at 100 mL/kg per 24 h. Calculation of fluid requirements based upon BSA (1.500 mL/mm^2 per 24 h) will vary only slightly. Fluid for maintenance is generally provided as 5% dextrose in 1/4 or 1/3 normal saline. For short-term intravenous therapy, sodium 5 meq/kg per day and potassium 2 meq/kg per day will satisfy the daily need. Fluid and electrolyte losses secondary to protracted vomiting or diarrhea are corrected by modifying this formula according to the measured losses. In the infant the normal serum osmolarity is between 280 and 290 mO/liter. However, the immature kidney is unable to concentrate efficiently, and therefore, it is not always possible to utilize the urine osmolarity as a guideline for fluid replacement in neonates. For example, even in the dehydrated infant, the urine osmolarity rarely exceeds 400 mO/liter. Hyperosmolarity is particularly hazardous in the neonate and can result in intracranial hemorrhage. Therefore, the administration of hypertonic solutions must be carefully monitored.

Whatever the formula used to calculated fluid replacement for the infant or small child, there is no substitute for collecting and analyzing fluid losses and replacing the depleted constituents precisely.

Acid-Base Equilibrium. Measurement of arterial blood gases permits the assessment of the status of alveolar ventilation and acid-base equilibrium. Just as in adults, these data are essential during the resuscitation of critically ill children and constitute an integral part of intraoperative and postoperative monitoring. Significant elevations of arterial P_{CO2} usually indicate the need for endotracheal intubation and mechanical ventilation. Impaired arterial oxygenation as indicated by reduced Pa_{O2} may be a consequence of compromised ventilation-perfusion secondary to pulmonary parenchymal disease or right-to-left shunting. From measurement of serum pH and P_{CO2}, base deficits can be determined utilizing the standard Siggard-Anderson curve nomogram. Generally half the calculated requirement for sodium bicarbonate is administered, after which blood-gas measurements are repeated and then additional corrections are carried out. For infants requiring extensive metabolic manipulation such as those with congenital diaphragmatic hernia, all intravenous sodium solution should be as sodium bicarbonate. This maximizes the amount of buffer provided while restricting sodium ion administration.

Fluid and electrolyte losses from the upper gastrointestinal tract in infants are generally equivalent to 0.45 N saline solution. Adjustments for ileostomy or biliary losses are made when these are significant by analyzing the individual drainage.

Blood Volume and Blood Replacement. A useful guideline for estimation of blood volume for the infant is 85 mL/kg of body weight. When whole blood is utilized, the transfusion requirement is calculated as 10 mL/kg, which roughly is equivalent to a 500-mL unit transfusion for a 70-kg adult. Replacement with packed cells is calculated at 5 mL/kg. In the child, coagulation deficiencies may rapidly assume clinical significance after extensive blood transfusion. It is advisable to alternate whole blood transfusion with the administration of fresh frozen plasma in order to replace the labile clotting factors.

Hyperalimentation and Nutrition. The physiologic nutritional demands imposed upon the growing infant are well recognized. When these are compounded by illness and the necessity to repair tissue and heal surgical wounds, the risks of protein calorie malnutrition are considerable. Thus, parenteral nutritional support has been integrated into the management of infants and children with surgical illness. When the gastrointestinal tract is not usable because of mechanical, ischemic, or inflammatory disorders, several options for nutritional support are available. Techniques for delivering calories by a central or a peripheral venous route have been refined to the stage that the caloric needs of all patients can be satisfied.

The original technique of parenteral nutrition made use of an indwelling central venous catheter for delivery of the nutritional substrate. The principle involved the provision of a source of calories (hypertonic glucose) in combination with the source of nitrogen (protein hydrolysate or amino acid solution). Essential fatty acid supplements, minerals, and vitamins are provided in the infusate, and long-term growth can be sustained even in the rapidly developing surgical infant. The risks of this technique are not trivial and include sepsis, caval thrombosis, pneumothorax, hydrothorax, and hypertonic crisis. For this reason alternatives to central venous alimentation have been developed. Peripheral alimentation, utilizing less concentrated but greater volume of solutions, in combination with intravenous lipid supplements has eliminated the need for central alimentation in many patients. In some centers peripheral alimentation techniques are employed almost exclusively. Solutions containing intravenous lipid preparations are administered by "piggybacking" this solution into the intravenous line. Alternatively, infusions or intravenous fat can be administered through a fine-gauge needle inserted into a peripheral vein. The infusion of all solutions utilized for alimentation and indeed any intravenous solution in an infant or small child is controlled by a properly alarmed, constant infusion pump. To prevent the development of trace metal deficiencies, supplementary copper, zinc, and iron are provided to patients receiving long-term parenteral nutritional support.

By utilizing these techniques, positive nitrogen balance can be accomplished for even premature infants. Refinements and advances in the techniques of parenteral nutrition have had an enormous impact on the survival of pediatric surgical patients.

Thermoregulation. Infants or children compromised by disease are extremely thermolabile. Premature infants are particularly susceptible to changes in environmental temperature. Because they are unable to shiver and lack stores of fat, their potential for thermogenesis is impaired. Since these patients lack adaptive mechanisms to cope with the

environment, the environment must be regulated. Attention to heat conservation during transport of the infant to and from the operating room is essential. Transport units incorporating overhead radiant heating which is servo-controlled by a skin temperature probe is recommended. When this is not available, the simple expedient of wrapping the infant in aluminum foil during transportation will diminish radiant heat loss. In the operating room the ambient temperature should approach thermal neutrality for the patient. This is approximately 21°C (73°F). Supplementary heat is provided by means of direct warming lights during positioning and endotracheal intubation when much of the body surface is exposed. Warmed solutions are utilized for "prepping" the operative field. Excessive solutions must be avoided to minimize evaporation with concomitant body surface cooling. Irrigating solutions employed during surgery should be warmed. Intraoperative temperature is monitored by placement of a thermistor probe in the axilla or esophagus.

Lesions of the Neck

Cystic Hygroma

Etiology and Pathology. Cystic hygroma occurs as a result of sequestration or obstruction of developing lymph vessels. The majority of lesions occur in the neck posterior to the sternocleidomastoid muscle. Other sites of involvement are the axilla, groin, and mediastinum. The term "hygroma" implies an endothelial-lined fluid-filled sac. The cysts may be unilocular, but more often multiple cysts permeate the surrounding structures and distort the local anatomy. Adjacent connective tissue often shows extensive lymphocytic infiltration. The mass may be apparent at birth or may appear and enlarge rapidly in the early weeks or months of life. Occasionally cystic hygromas contain nests of vascular tissue. A particularly troublesome variant of cystic hygroma is that which involves the tongue, floor of the mouth, and structures deep in the neck.

Infection within the cysts can cause rapid enlargement which may result in airway compromise. Rarely, it may be necessary to carry out percutaneous aspiration of an infected cyst to relieve respiratory distress.

Treatment. Surgical excision offers the best chance of cure. Total excision is often impractical because of the extent of hygroma and its proximity and interdigitation with adjacent nerves and vascular structures. Because this lesion is benign, regional ablative surgery is not indicated. Rather, repeated partial excision of residual hygroma is advised. Some hygromatous tissue is left behind when cysts are unroofed and partially excised. However, this is preferred to sacrificing branches of the facial nerve or other crucial structures. Postoperative wound drainage is important and is best accomplished by closed-suction technique. Occasionally fluid accumulates beneath the surgically created flaps in the area from which the hygroma was excised. Multiple needle aspirations may be required to eliminate accumulated fluid.

Thyroglossal Duct Remnants

Pathology and Clinical Manifestations. The thyroid gland originates as a diverticulum off the foregut beginning at the base of the tongue in the foramen cecum and passes downward and forward through the hyoid bone to the normal anatomic position of the

thyroid gland. Thyroid rest tissue left behind in the migration may persist and present in the midline of the neck as a thyroglossal duct cyst. The mass is usually discovered in the two- to four-year-old child when the baby fat subsides and irregularities in the neck are more readily apparent. Almost always the cyst is encountered in the midline just at or below the level of the hyoid bone. Occasionally regional infection results in distortion of the anatomy, and lateral displacement of the cyst can occur which may lead to diagnostic confusion. Most thyroglossal duct cysts are asymptomatic. Occasionally they become infected and present as a tender swelling or abscess in the midneck. The differential diagnosis includes submental lymphadenopathy and midline dermoid cysts. Rarely, midline ectopic thyroid tissue masquerades as a thyroglossal duct cyst. In this circumstance, the ectopic thyroid may represent the patient's only thyroid tissue. Therefore, if there is any question regarding the diagnosis or if the thyroid gland cannot be palpated in its normal anatomic position, it is advisable to obtain a nuclear scan to confirm the presence of a normal thyroid gland.

Treatment. Patients with infected thyroglossal duct cysts should be treated with antibiotics; when an abscess is present, drainage is necessary. It is never advisable to carry out surgical excision through inflamed tissues. Removal of the cyst and thyroglossal duct including the central portion of the hyoid bone is curative.

Branchial Cleft Anomalies

Branchial clefts are those embryological grooves that ultimately evolve into the lower part of the face and neck. Most congenital fistulas result from a malformation or persistence of the second branchial cleft. When the fistula is complete, it extends from the anterior border of the sternocleidomastoid muscle superiorly, then inward between the bifurcation of the carotid artery, entering the posterolateral pharynx just below the tonsillar fossa.

Diagnosis and Treatment. These children usually present with a painless nodule at the anterior margin of the sternocleidomastoid muscle. Often there is clear fluid draining from the tract when the ostium opens externally. Incomplete sinus tracts frequently contain small bits of cartilaginous tissue. The treatment is surgical, and complete removal of the cyst and tract is necessary for cure. Dissection of the sinus tract is facilitated by passing a fine lacrimal duct probe through the external opening into the tract and utilizing this as a guide for dissection. A series of two or sometimes three small "stair-step" incisions are preferred to a long oblique incision in the neck which is cosmetically unacceptable.

Respiratory System

Subglottic Stenosis

Congenital narrowing of the subglottic region usually occurs several millimeters below the level of the vocal cords. The hallmark of upper airway obstruction is inspiratory and expiratory stridor; congenital obstruction is usually a result of thickened subglottic tissues or abnormal cartilaginous overgrowth. Acquired stenosis is seen more frequently as infants are being sustained by ventilatory support requiring endotracheal intubation. Most patients with subglottic narrowing of the acquired type will outgrow the problem as the larynx enlarges. The occasional patient will require periodic dilatations to maintain airway patency.

Tracheoplasty employing prosthetic materials for stenting the upper airway is reserved for the most severe stenoses which do not respond to dilatation.

Subglottic Hemangioma

Abnormalities of blood vessels with resultant hemangioma can occur in the subglottic area and may narrow the airway, resulting in stridor. These lesions can often be demonstrated on a high-kilovolt neck film. There is some hazard to instrumentation of the airway when a subglottic hemangioma is present. In some patients tracheostomy may be required, but the natural history of hemangioma is to increase in size during the first few months, after which there is a gradual involution and disappearance of the lesion. When airway obstruction is critical, involution of the hemangioma can sometimes be hastened by the systemic administration of corticosteroids in combination with radiation therapy to the subglottic area.

Congenital Diaphragmatic Hernia (Bochdalek)

Pathology. During the formation of the diaphragm, the pleural and coelomic cavities remain in continuity via the pleuroperitoneal canal. This posterior lateral communication is the last to be closed by the developing diaphragm. Failure of diaphragmatic development leaves a posterior lateral defect known as a Bochdalek hernia. This anomaly is more commonly encountered on the left.

With incomplete development of the posterior diaphragm, bowel fills the chest cavity during embryonic life. Consequently the abdominal cavity remains small and undeveloped. The presence of intestine in the chest interferes with the development of the lung and prevents its expansion after birth. Following delivery, as the bowel fills with air, cardiorespiratory dynamics are further compromised as the bowel distends and the mediastinum is shifted. The outcome is catastrophic unless the situation is recognized and corrected. Any newborn infant in respiratory distress who shows a scaphoid abdomen should be suspected of having a congenital diaphragmatic hernia. Chest x-ray will confirm the diagnosis.

Treatment. While diaphragmatic hernia is acknowledged to be the most urgent surgical emergency in the newborn infant, if does not follow that the infants presenting earliest do best. In fact, the opposite is true. It has been observed in many centers that infants with diaphragmatic hernia recognized after the first 24 hours enjoy a better prognosis than those presenting with respiratory distress immediately after birth or within the first day of life. To account for this apparent paradox it is assumed that the more severely involved infants present earliest, while those babies who remain compensated throughout the first days of life have lesser degrees of pulmonary hypoplasia. The most severely involved infants succumb within hours of operative correction. Another group of infants has been identified who seem to do well immediately after surgery and for hours and even days. After this period of apparent well-being, there is clinical deterioration characterized by hypoxia and cyanosis presumably a result of right-to-left shunting and persistent fetal circulation. Pharmacologic support with vasopressors and pulmonary vasodilators may result in temporary improvement, but the majority of these infants succumb to anoxia and acidosis despite maximal ventilatory support.

Some severely compromised infants may benefit from a brief period of resuscitation with sodium bicarbonate and mechanical ventilation with 100% oxygen before the surgical correction. Improved survival appears to correlate with the preoperative correction of acidosis, hypercarbia, and hypoxia. In those infants in whom ventilation with 100% oxygen fails to result in an elevation of P_{O2} above 100 torr, the outlook is grave. For those infants in whom a P_{O2} of greater than 100 torr with a P_{CO2} less than 40 can be achieved, the outcome after surgery is more favorable. This correlation undoubtedly reflects the severe degree of pulmonary hypoplasia in the former group.

Operative repair can be accomplished by either a transthoracic or an abdominal approach. Diaphragmatic repair is facilitated by the former, but there are advantages to approaching the lesions through the abdomen, and this is generally recommended. Through a subcostal incision the abdominal viscera are withdrawn from the chest, exposing the defect in the diaphragm. The anterior margin of the diaphragm is generally apparent, while the posterior muscular rim is usually somewhat attenuated. Careful dissection of the posterior rim of the diaphragm, which is usually covered by a layer of peritoneum continuous with the parietal pleura, will allow this posterior muscle component to be unrolled so that it can be sutured to the anterior diaphragmatic muscle. A single layer of nonabsorbable suture is used to close the defect. In rare instances, primary repair will be impossible, and it will be necessary to employ a patch of prosthetic material. Just before the diaphragmatic repair is completed, a chest tube is positioned in the thoracic cavity and placed to gentle suction.

It is advisable to place a chest tube on the contralateral side because of the relatively high incidence of pneumothorax opposite the side of the diaphragmatic hernia. With a chest tube in place, a life-threatening pneumothorax is avoided.

It may be impossible to accomplish an anatomic closure of the abdominal wall after return of the viscera to the abdominal cavity. In this circumstance only the skin is closed, creating a ventral hernia. By this maneuver, pressure on the diaphragm and inferior vena cava is relieved. The ventral hernia is repaired 10 days to 2 weeks later, after the abdominal cavity has expanded to accommodate the intestine. In spite of improvement in neonatal pulmonary support, the mortality from diaphragmatic hernia, especially in infants symptomatic immediately after birth, remains extremely high. Bartlett and others have demonstrated that infants with potential respiratory failure can be supported effectively by extracorporeal membrane oxygenation (ECMO). Infants with diaphragmatic hernia in whom death was inevitable have been successfully supported by this technique. Currently, veno-arterial bypass established by cannulating the right atrium through the jugular vein and the aortic arch through the right common carotid artery has been proved feasible in these infants. Respiratory failure secondary to diaphragmatic hernia has become one of the important indications for pulmonary support by ECMO.

Congenital Lobar Emphysema

Hyperexpansion of one or more lobes of the lung can result in a life-threatening respiratory crisis. Inspired air is trapped in the affected lobe and results in compression of the adjacent lung and displacement of the mediastinum. The right upper, right middle, and left upper lobes are most frequently involved. In 10 percent of cases there is bilobar involvement. When overdistension of the lobe progresses rapidly in the newborn infant, dyspnea, cyanosis,

and severe respiratory compromise can result. More commonly, the overdistension of the lobe is gradual, and symptoms do not occur until later in life. It is presumed that the affected lobe does not deflate normally because of a deficiency of cartilage in the bronchus supplying that lobe. However, histologic examination of resected specimens rarely identifies a specific bronchial abnormality.

In some patients the condition may be self-limiting or stable, and surgery is not required. In others, lobar emphysema may pose a true surgical emergency, and the affected lobe must be removed promptly to relieve respiratory symptoms.

Pulmonary Sequestration

Accessory lung tissue may be present, either completely separate from adjacent lung parenchyma (extralobar sequestration) or invested within the visceral pleura of the adjoining normal lung tissue (intralobar sequestration). The former condition occurs most commonly on the left side of the chest adjacent to the diaphragm. The accessory lobe is not attached to the adjacent lung and is covered by a smooth pleural surface. There is no bronchial connection, and the blood supply to the anomalous pulmonary tissue arises from the systemic circulation. Extralobar sequestrations are asymptomatic, and this malformation is usually discovered on a routine chest film. Arteriography is helpful in demonstrating the systemic arterial supply. Elective excision is advisable.

Intralobar sequestration occurs in intimate association with normal lung parenchyma, usually in the lower lobes and more commonly on the left. Unlike the asymptomatic extralobar variety, intralobar sequestrations are often associated with cough, hemoptysis, and recurrent pulmonary infection. There is no connection with the bronchus, but the sequestration may contain air which enters from adjacent alveoli. Resection of the sequestration often necessitates a lobectomy, though segmental resection may sometimes suffice. The blood supply to the sequestrated tissue arises from the systemic circulation, often the aorta below the diaphragm; therefore, preoperative arteriography is strongly advised.

Bronchiectasis

Bronchiectasis is rare in children but can occur as a late sequela of pneumonia. Children with cystic fibrosis often develop bronchiectasis as a complication of underlying lung disease. Bronchiectasis may also result from infection secondary to a neglected bronchial foreign body. Symptoms include chronic cough often productive of purulent secretions. Recurrent pulmonary infection and hemoptysis are not uncommon. The diagnosis is suggested on chest x-ray, and the saccular or fusiform distortion of peripheral bronchi can be demonstrated by bronchography. Lobectomy or segmental resection is indicated if the disease is localized to a single lobe or portion of a lobe.

Foreign Bodies

Foreign objects aspirated into the airway represent a major cause of respiratory difficulty in toddlers. Peanut aspiration is common, and severe lipoid pneumonia may result unless the offending nut fragment is extracted.

A history of choking while eating, often when a child is playing or laughing, should alert one to the possibility of foreign body aspiration. The coughing may then resolve, and the only sign of foreign body in the bronchus may be a unilateral wheeze. Many of these children are treated for "asthma" without response before the clinician considers the diagnosis of aspirated foreign body. A chest x-ray often reveals hyperaeration (obstructive emphysema) involving the whole lung or localized to a single lobe. Fluoroscopy of the chest will often reveal air trapping and a swing of the mediastinum toward the side of the foreign body during expiration. In neglected cases, atelectasis ensues, and bronchiectasis is a late consequence of an unrecognized bronchial foreign body. Although some foreign bodies are expelled by treatment with pulmonary physiotherapy, endoscopic removal under general anesthesia is recommended. Occasionally it is necessary to perform endoscopic procedures under fluoroscopic control. By this technique a Fogarty catheter is inserted past the foreign body, the balloon inflated, and the foreign material extracted.

Pneumothorax, Pneumomediastinum

The improved salvage of premature infants with respiratory distress syndrome has inevitably resulted in long-term maintenance on mechanical ventilation. Diminished compliance of the lungs combined with positive-pressure ventilation results in an appreciable incidence of pneumomediastinum and pneumothorax. The development of pulmonary interstitial emphysema (air surrounding the minor bronchi) precedes dissection of the air extrapleurally into the mediastinum or the pleural cavity. Pneumomediastinum is not usually clinically significant, but may contribute to respiratory distress and require needle aspiration. This can be accomplished safely by inserting a needle into the mediastinum beneath the xiphoid. Pneumothorax in neonates is almost always under tension and thus life-threatening. Sudden deterioration of the infant's clinical status with a decrease in arterial oxygen saturation and hypercarbia often signals a pneumothorax. Chest x-ray is confirmatory, and the pneumothorax must be aspirated as an emergency. Even if immediate relief of pneumothorax is obtained by needle aspiration, it is prudent to insert a small chest tube for a few days.

Esophagus

Tracheoesophageal Fistula and Esophageal Atresia

Esophageal Atresia with Tracheoesophageal Fistula

In 1939, Ladd in Boston and Leven in Minneapolis, working independently, successfully managed two infants with esophageal atresia and tracheoesophageal fistula. Both performed gastrostomy followed later by fistula ligation. The children survived; each ultimately had esophageal reconstruction with an antethoracic skin tube. In 1941, Dr Cameron Haight in Ann Arbor performed the first successful primary anastomosis for esophageal atresia.

Gray and Skandalakis offer a lucid description of the formation of the esophagus and trachea in the embryo. The esophagus and trachea are first recognized as a ventral diverticulum of the foregut about 22 or 23 days after fertilization. As the diverticulum elongates, a proliferation of endodermal cells appears on its lateral walls. In 1931, Rosenthal first observed that these cell masses become ridges of tissue that ultimately divide the foregut

into tracheal and esophageal channels. The division into separate tubes is completed between 34 to 36 days, at which time the submucosal and muscular layers of both esophagus and trachea are apparent.

Among esophageal malformations, 85 percent of the defects occur as a blind-ending upper esophageal segment with the lower portion of the esophagus connected to the trachea (Gross-Vogt type C). The esophageal fistula usually joins the trachea at or just above the tracheal bifurcation, admitting inspired air into the stomach and, in a retrograde fashion, gastric juice into the lungs.

Associated anomalies are common and often are the most significant factor influencing survival. In nearly 20 percent of the babies born with esophageal atresia, some variant of congenital heart disease occurs, while imperforate anus is found in about 12 percent.

Clinical Manifestations. The earliest and most obvious clinical sign of esophageal atresia is regurgitation of saliva or of the first offered feeding. Aspiration of a feeding is often followed by choking or coughing. Abdominal distension is a prominent feature, occurring as inspired air is transmitted through the fistula into the stomach. Regurgitated gastric juice passes through the fistula and into the trachea and lungs, leading to chemical pneumonia. The pulmonary problems are compounded by atelectasis, which occurs from diaphragmatic elevation secondary to gastric distension. The diagnosis is not obvious at the initial newborn examination by the physician unless an attempt is made to pass a tube into the stomach. Nurses who are feeding the baby and observing behavioral patterns such as the accumulation of mucus or saliva often make the early diagnosis.

A contrast x-ray study confirms the diagnosis. Contrast medium instilled into the pouch outlines the blind upper esophagus. Tracheal air enters the stomach via the fistula and establishes the presence of a tracheoesophageal communication on the x-ray. The condition of the lungs and the existence of pneumonia or atelectasis are also essential information.

Treatment. With the diagnosis secure, the following measures should be instituted immediately: (1) infant warmer; (2) 30° head-up position; (3) route for intravenous therapy; (4) antibiotic treatment (even if pneumonia is not yet clinically manifest); and (5) sump catheter suction in the upper pouch. Gastrostomy is a useful adjunct in the management of many babies before operative repair of the esophagus is undertaken.

In 1962, Waterston et al developed risk categories for infants with esophageal atresia and tracheoesophageal fistula. They classified the infants as follows:

A. Birthweight over 5.5 lb and otherwise well.

B. 1. Birthweight 4 to 5.5 lb and well.
2. Or higher birthweight, moderate pneumonia, and other congenital anomaly.

C. 1. Birthweight under 4 lb.
2. Or higher birthweight but severe pneumonia and severe congenital anomaly.

The babies were categorized this way in order to predict more accurately the outcome that could be expected. In our institution, a separate treatment plan has been devised for each group.

Category A: Prompt Surgical Correction. If the infant is full-term, has no significant pneumonia, and shows no other major congenital anomalies, primary repair can be undertaken safely.

The operative technique for primary repair is depicted. The operation for primary repair of esophageal atresia and tracheoesophageal fistula has changed very little since its original description by Cameron Haight. Although the transpleural approach may reduce the operating time, most workers now agree that the retropleural approach is desirable. Exposure with this method is perfectly adequate, and the protection of the lung by maintaining its pleural envelope has a salutary postoperative effect. More important, an anastomotic leak will not communicate with the pleural cavity and can be drained directly from the mediastinum posteriorly with decreased morbidity or mortality.

Livaditis showed that incising the muscle of the upper pouch circumferentially produced remarkable lengthening without compromising blood supply. This approach is useful if the distance between the two pouches precludes anastomosis without producing undue tension.

Category B: Surgical Repair Following Short-Term Delay. If the infant has pneumonia or weighs less than 5 lb, surgery is deferred. A gastrostomy is always performed, sometimes under local anesthesia. Time is thus provided for stabilization and medical management of the pulmonary status. This method of management is useful for only a limited period, because reflux into the tracheobronchial tree via the tracheoesophageal fistula may occur in spite of gastric decompression through the gastrostomy. Repair is carried out as described above when the infant's condition stabilizes and the risk of surgery has been reduced.

Category C: Staged Repair. In babies with serious coexisting anomalies, prematurity, low birth weight, or persistent pulmonary disease, survival is limited. The possibility of diminishing the mortality rate in compromised infants by staging the operative procedure has received considerable study. Although staged repair is used much less frequently today than a delayed primary repair, the approach has merit in selected patients, but staging of the operation has by no means led to universally acceptable results. The outstanding work of Dudrick and his coworkers in the field of parenteral nutrition has added a new dimension in the care of these difficult babies. It is now possible to provide nutritional support indefinitely for newborn infants by a central venous or even peripheral intravenous routes. This advance, coupled with the holding pattern provided by suction of the upper pouch and gastrostomy drainage, makes it possible to maintain infants with esophageal atresia and tracheoesophageal fistula indefinitely while growth and weight gain are achieved, pulmonary status is cleared, and other congenital anomalies are studied and corrected. Correction of this form of esophageal anomaly leads to a satisfactory outcome with nearly normal esophageal function in most patients.

Complications. Although once considered fatal, a leak at the anastomosis can usually be dealt with satisfactorily, particularly if the pleural envelope has been maintained and drainage accomplished from the mediastinum via the retropleural route. Improved management of infection, nutrition, and respiratory support in infants has contributed to success in handling anastomotic leaks.

The mediastinal infection seen with esophageal disruption may cause reformation of the tracheoesophageal fistula. This complication requires another operation to divide the tracheoesophageal fistula.

In the past decade it has become apparent that the necessary force applied to the esophagus to complete the anastomosis may alter the anatomy of the gastroesophageal junction, leading to gastroesophageal reflux. The clinical manifestations of this reflux are identical to those seen in other infants with primary gastroesophageal reflux.

Isolated Esophageal Atresia

Among those babies born with esophageal anomalies, 8 percent have isolated esophageal atresia. Characteristically, infants with isolated esophageal atresia present with a scaphoid abdomen, since the gastrointestinal tract is devoid of air. The x-ray finding of a blind upper pouch and the absence of air below the diaphragm is pathognomonic of isolated esophageal atresia without fistula.

Prompt esophagostomy with the upper esophageal pouch brought to the skin of the left side of the neck allows drainage of saliva and prevents aspiration. A gastrostomy is performed and serves for feeding in the early months of life. Esophageal replacement with colon or gastric tube is then recommended at a year of age. Some authors have shown that the esophageal ends can be dilated over a period of several months, allowing end-to-end union and avoiding esophageal replacement.

Isolated (H-Type) Tracheoesophageal Fistula

In rare instances (3 percent), an isolated congenital fistula connecting the trachea to the esophagus may exist. In this anomaly both the trachea and the esophagus are otherwise normal, with no narrowing or obstruction. Infants with this condition seem to swallow normally. The clinical features are subtle; weeks or months may elapse before a correct diagnosis is made. The presence of an H-type tracheoesophageal fistula is suggested by the following triad of symptoms: (1) choking when feeding, (2) gaseous distension of the bowel, and (3) recurrent aspiration pneumonia. Diagnosis can usually be confirmed by cine contrast x-ray studies or by bronchoesophagoscopy using the recently available fiberoptic lens system. Definitive treatment consists of dividing the fistula. Surgical closure of the esophagus and trachea must be meticulous, and encroachment on the lumen of the trachea voided. The fistula is usually accessible to surgical repair through an incision just above the clavicle.

Corrosive Injury of the Esophagus

Commercial cleaners stored in areas accessible to the toddler are commonly swallowed. These agents often contain a strong alkali which is hygroscopic and becomes

firmly attached to the moist mucosal surface of the esophagus. The burn thus sustained coagulates protein and can involve the entire thickness of the esophagus. Children suspected of having swallowed corrosive materials should be admitted to the hospital and studied by esophagoscopy within 24 hours of injury. A burn will be seen as a whitish coagulum on the surface mucosa, surrounded by an area of hyperemia. If the esophageal injury is circumferential, it has been our practice to perform a gastrostomy for feeding, and to insert a string for subsequent dilatation.

Steroids seem to decrease the mucosal scarring and are therefore administered for 6 weeks. Antibiotics are used routinely for 3 weeks. After this interval, esophageal dilatation is begun. Dilatations are continued as often as twice weekly until either the stricture is resolved or it becomes apparent that esophageal substitution will be required. This latter decision is deferred for at least 6 to 12 months until healing is complete and the resulting esophageal injury can be fully assessed.

Esophageal Substitution

Esophageal substitution is required in children for two major conditions: severe esophageal strictures and isolated esophageal atresia. The colon has been the most widely used organ for esophageal substitution, reaching easily into the neck on the marginal artery of Drummond. It can be placed in a substernal tunnel or in the left side of the chest behind the lung root. Since the colon acts as an aperistaltic conduit, antiperistaltic and isoperistaltic segments function equally well. An alternative method of esophageal substitution gaining popularity is the reversed gastric tube. This is fashioned from a flap cut from the greater curvature of the stomach, with a vascular pedicle based on the left gastroepiploic artery. The results of both these methods of esophageal substitution are satisfactory. Children so treated are able to maintain normal growth and development, and the long-term complications are manageable.

Gastroesophageal Reflux

In 1950, Berenberg and Neuhauser described a group of babies who vomited excessively and were demonstrated to have unimpeded gastroesophageal reflux. Their term, "chalasia" (relaxation of the cardia) became widely accepted, and the upright position that these workers advocated became the standard mode of therapy. A certain percentage of infants with "chalasia" were not controlled by the upright propping technique; in these babies persistent gastroesophageal reflux caused serious consequences in growth and development and in the respiratory tract. In the past two decades, pernicious gastroesophageal reflux, occurring without demonstrable hiatal hernia, has become recognized as a unique entity found in infants during their first year of life. Surprisingly, in this particular age group, the incidence of esophagitis and stricture is infrequent.

The foregoing clinical picture is clearly separate from that of children older than 1 year who present with more typical hiatal hernia and gastroesophageal reflux. In this group of children, who range in age from 2 years through adolescence, esophagitis with classic symptoms of substernal discomfort, progressing to stricture formation is a more characteristic end point. In some children with gastroesophageal reflux, documentation has appeared that relates the presence of reflux to a variety of clinical presentations including seizures,

hyperirritability, peculiar posturing, dystonia and developmental retardation, protein-losing enteropathy, asthma, and chronic respiratory disease. Furthermore, the association of gastroesophageal reflux following repair of esophageal atresia is reported with increasing frequency.

Clinical Manifestations. History of repeated episodes of vomiting in an infant is the clearest indication of gastroesophageal reflux after obvious anatomic obstruction at or beyond the pylorus has been excluded. When the vomiting is associated with failure of normal development or chronic respiratory symptoms, the likelihood of pernicious gastroesophageal reflux is increased.

A contrast esophagogram will demonstrate reflux in the majority of infants and children so affected. Darling has characterized and quantified types of esophageal reflux in infants and children as demonstrated by esophagogram. However, barium swallow remains an imperfect tool for diagnosis. Miniaturized equipment is now available to allow constant monitoring of esophageal pH even in infants. Analysis of these recordings can show with accuracy the frequency and character of the reflux episodes. Esophagoscopy coupled with biopsy has proved useful in assessing the presence or extent of esophagitis.

Treatment. After definitive diagnosis most patients are treated initially by conservative means. In the infant, this means maintenance of the upright position. Thickening the formula with rice cereal also decreases vomiting. In older patients medical measures include antacids, dietary restrictions, and elevation of the head of the bed as in adult patients. Exceptions to a trial of medical therapy after establishing the diagnosis of gastroesophageal reflux in infants and children are as follows:

1. Life-threatening apneic spells related to the reflux.

2. Congenital displacement of a major portion of the stomach in the chest.

3. Significant esophagitis unaffected by medical therapy.

4. Established stricture.

5. Chronic pulmonary changes.

Most infants and children respond favorably to one of the standard antireflux procedures. The Nissen fundoplication is probably the most widely used operation in younger patients, but partial wraps (Thal), Belsey Mark IV, and Boerema gastropexy all have been reported with success.

Gastrointestinal Tract

Pyloric Stenosis

Clinical Manifestations. The typical infant with hypertrophic pyloric stenosis is 7 weeks of age, male, and the first-born child. Nonbilious vomiting, becoming increasingly projectile in nature, occurs over several days to weeks. Eventually the infant will not even

hold down water and becomes severely dehydrated, showing a metabolic alkalosis and severe depletion of potassium and chloride ions. Potassium deficits may not be apparent until late stages since the serum potassium is maintained even after severe losses. Serum pH is high; urine pH is initially high but eventually drops as severe potassium deficit leaves only hydrogen ions for exchange with sodium ions in the distal tubule of the kidney.

The diagnosis of pyloric stenosis can usually be made on physical examination, the typical "olive" being palpable in the right upper quadrant. On occasion vigorous peristaltic waves can be seen passing from left to right across the epigastrium. Rarely, ultrasound or limited upper gastrointestinal series, using minimal amounts of barium, may be needed to confirm the diagnosis.

Treatment. Reversal of electrolyte abnormalities and metabolic alkalosis is essential before operation. For severe depletion a normal saline solution with added potassium (2 to 4 meq/kg over 18 to 24 h) at volumes sufficient to reverse dehydration and establish good urine flow (1 to 2 mL/kg per h) will be necessary. After resuscitation, a Fredet-Ramstedt pyloromyolotomy is performed. Postoperatively, intravenous feedings are continued for several hours, after which small frequent feedings of dilute formula are offered with gradual increase to bolus feeds of full-strength formula. The infant can usually be discharged within 48 hours of surgery.

Pneumoperitoneum

Pneumoperitoneum in the neonate is a surgical emergency. The commonest cause at this time is probably perforation of gangrenous bowel of necrotizing enterocolitis (see later). Also included in the differential diagnosis are idiopathic gastric perforation, perforation of the colon in Hirschprung's disease, and occasionally breakthrough of mediastinal and retroperitoneal air in the infant with severe respiratory distress syndrome requiring high ventilator pressures.

Pneumoperitoneum itself can cause respiratory embarrassment by elevating the diaphragm and compromising lung volume. This can be alleviated by needle aspiration of the abdomen in the epigastrium before definitive surgical treatment of the underlying condition. If the diagnosis of pneumoperitoneum secondary to pulmonary air leaks can be established nonoperatively (such an infant would manifest no signs of peritonitis), repeated aspiration may be performed until ventilator pressures can be lowered.

Gastrostomy

The performance of a gastrostomy in an infant avoids the potential complications of an indwelling nasogastric tube, namely respiratory difficulty secondary to gastroesophageal reflux and aspiration, and blockage of the nares (infants are obligate nose breathers). Feeding by gastrostomy may be necessary for prolonged periods in small or sick infants and infants with orofacial anomalies or swallowing deficits. Such feedings can be performed safely by parents at home.

A Stamm gastrostomy is performed through a tiny vertical left upper quadrant incision. For emergency gastrostomies a Malecot catheter is inserted into the stomach and tied with a

double purse-string suture and brought out through the wound. If prolonged use of the gastrostomy is anticipated, the catheter is brought out through a separate stab wound. It is important to suture the stomach to the abdominal wall to avoid intraperitoneal soilage by gastric contents. After the gastrostomy is no longer required, the Malecot catheter is removed and usually the stab wound will close spontaneously in a few days. Formal closure may be required if the gastrostomy has been in place for many months.

Intestinal Obstruction in the Newborn

Bilious vomiting is the most common manifestation of intestinal obstruction in the newborn. High obstruction such as duodenal and jejunal atresia produces early vomiting in the first 24 hours of life. The later the onset of vomiting, the lower the obstruction is likely to be. Failure of passage of meconium is characteristic of lower ileal and colonic obstruction. The degree of abdominal distension is also correlated roughly with the level of obstruction. Distended loops of bowel, the number corresponding to the level of the obstruction, and air-fluid levels on upright abdominal films are seen commonly and may be the only diagnostic studies necessary before surgery. Usually, 30 to 40 mL of air injected into the stomach is a satisfactory "contrast" material for high obstructions, and barium given from above is rarely necessary. Contrast enema may show a "micro" or unused colon in low obstruction.

Duodenal Malformations

Duodenal obstruction may be complete, as in duodenal atresia, or partial, as in duodenal web or stenosis, annular pancreas, and malrotation of the midgut. Anomalous entry of the common bile duct and pancreatic duct may be associated, requiring caution when anomalies are dealt with surgically by local plastic procedures on the duodenum. Although bile vomiting is most often seen, entry of the bile duct distal to the site of obstruction may occur. In this case vomitus contains only clear gastric contents. The "double bubble" seen on an air-contrast upper gastrointestinal series is characteristic of duodenal atresia, with small amounts of air seen distally if obstruction is incomplete.

The duodenum can be adequately decompressed via nasogastric tube so that surgical correction is relatively nonurgent. This is of importance because one-third of infants with duodenal obstruction have Trisomy-21. Time is needed to look for other anomalies (such as cardiac) in these infants as well as to establish the chromosomal aberration by karyotyping. Surgery should not be deferred if malrotation is the cause of duodenal obstruction or if malrotation is present with intrinsic duodenal obstruction.

Congenital obstruction occurs almost exclusively in the second portion of the duodenum. The surgical treatment of duodenal obstruction is adapted to the anatomic situation found on exploration. Atresia, stenosis, and annular pancreas are bypassed via duodenostomy or duodenojejunostomy, performing the anastomosis between the most dependent portion of the proximal duodenum and the distal bowel in end-to-end (duodenum) or end-to-side (jejunum) fashion. Webs can be excised through a vertical duodenal incision, the mucosa oversewn, and the duodenotomy closed horizontally. Gastrostomy is performed routinely.

Jejuno-Ileal Atresia

There is good evidence, both experimental and clinical, that intestinal atresia is the result of interruption of the vascular supply to the bowel at the fetal stage of development. Tying off the end arterial branches in the mesentery of experimental fetal animals will reproduce all the variations of obstruction seen in the human infant. Bowel deprived of its blood supply may form a web, stenosis, or single or multiple atresias. The bowel and its mesentery may reabsorb, leaving a characteristic V-shaped mesenteric defect. The length of intestine remaining may be quite short, which can result in prolonged feeding difficulties characteristic of the "short-gut" syndrome. The most severe form of jejuno-ileal atresia seen is the so-called Christmas-tree deformity in which a single artery (usually the ileocolic) is all that remains of the superior mesenteric artery, supplying, in retrograde fashion, a very short ileum which arranges itself in reversing spirals around its blood supply.

The clinical presentation of newborn infants with jejunal or ileal atresia is similar to that found in other types of intestinal obstruction. Bilious vomiting is characteristic, its onset related to the level of obstruction. Abdominal distension is progressive, being confined to the upper abdomen in high lesions, increasingly generalized the lower the obstruction. The number and distribution of obstructed loops on upright abdominal films offer some indication of the level of obstruction.

If the obstruction is distal or if substantial loss of intestinal length has occurred, there is insufficient length of ileum distal to the atresia to produce "succus entericus" in sufficient quantity to dilate the colon to its normal neonatal size. A contrast enema will therefore show a "microcolon". Since the proximal distended loops cannot be adequately decompressed by nasogastric tube from above, accumulation of gas and fluid may be sufficient to produce perforation with resulting nonsterile peritonitis. In instances of prenatal perforation, the resulting meconium peritonitis may lead to characteristic calcifications on abdominal films.

Surgical correction of small bowel atresia is urgent. Disparity in lumen size between the proximal distended bowel and distal collapsed bowel has led to a number of innovative techniques of anastomosis of the atretic bowel. These include (1) end-to-back technique, fish-mouthing the antimesenteric border of the distal loop; (2) tapering of the proximal distended loop to correspond to the distal loop with end-to-end anastomosis; (3) Bishop-Koop end-to-side union with exteriorization of the distal lumen; and rarely (4) Mikulicz exteriorization or (5) end ileostomies with delayed anastomosis.

Malrotation and Midgut Volvulus

Pathology. During fetal development the intestine elongates too rapidly to be accommodated in the abdominal cavity. Around the sixth week the midgut, supplied by the superior mesenteric artery, prolapses into the umbilical cord and remains until the tenth week. As the midgut returns to the abdominal cavity, the developing cecum and the duodenum undergo a counterclockwise 270° rotation around the superior mesenteric artery, the final C-loop of the duodenum, and the transverse and ascending colon tracing the path of the rotation. The duodenum then becomes fixed retroperitoneally in its third portion, emerging at the ligament of Treitz, and the cecum is fixed by peritoneal bands to the right lateral abdominal wall. In addition, the takeoff of the branches of the superior mesenteric artery becomes

elongated and fixed along a line extending from the epigastrium to the right lower quadrant. If rotation is incomplete, the cecum remains high and the duodenum becomes shaped like a corkscrew to the right of the superior mesenteric artery. The clinical consequences of these events are (1) the bands (Ladd's bands) attempting to fix the cecum to the abdominal wall straddle the duodenum and may obstruct it, and (2) the mesenteric takeoff remains confined to the epigastric region. The entire midgut and its vascular supply are therefore suspended on a narrow pedicle which may twist. This midgut volvulus results in high intestinal obstruction and midgut ischemia as the superior mesenteric blood flow is cut off.

Clinical Manifestations. Such an infant presents with irritability and bilious vomiting with later manifestations of vascular compromise such as blood stools. Abdominal signs are minimal early and may consist of mild upper distension and tenderness. As ischemia advances, erythema and edema of the abdominal wall and ultimately septic shock will signal gangrenous intestine and peritonitis. Abdominal films will show paucity of gas throughout the intestine with a few scattered air-fluid levels. A barium enema may show a displaced cecum, but an upper gastrointestinal series showing absent duodenal rotation makes a more reliable diagnosis.

Treatment. Early surgical intervention is mandatory if the ischemic process is to be reversed. The volvulus is untwisted counterclockwise followed by lysis of the bands between cecum and abdominal wall and between duodenum and terminal ileum to splay out the superior mesenteric artery and its branches. This procedure, originally described by Ladd, is still preferred. These maneuvers place the duodenum on the right and the cecum on the left of the abdomen. The appendix is removed. It is not necessary to place any holding sutures in cecum or duodenum if the bands have been fully lysed. When the viability of the midgut is in doubt, the volvulus is reduced and all compromised bowel returned to the abdomen. A "second look" 24 to 36 hours later will often show remarkable vascular recovery, and only irretrievably necrotic bowel is then resected.

Meconium Ileus

The bowel obstruction in meconium ileus is a result of impaction of meconium in the distal ileum. Such infants have cystic fibrosis, and the accompanying lack of pancreatic enzymes in the intestine contributes to the viscous nature of the meconium. Bile vomiting is a late feature of this condition, which is characterized by progressive abdominal distension, failure of passage of meconium, and an upright abdominal film showing enormously distended small bowel without air-fluid levels. Small bubbles of gas trapped in the inspissated meconium in the terminal ileum may produce a characteristic "ground-glass" appearance; a contrast enema will show a microcolon and terminal ileum filled with pellets of meconium. The fetal reaction to prenatal perforation causes intraabdominal calcifications to form, producing an eggshell pattern on plain abdominal x-rays. Uncomplicated meconium ileus, ie, that unassociated with perforation or vascular compromise of the distended ileum, can usually be resolved nonoperatively by a technique described by Noblett. Dilute Gastrografin is advanced through the colon under fluoroscopic control into the dilated portion of the ileum. Since Gastrografin acts partially by absorbing fluid from the interstitial fluid into the intestinal lumen, maintaining adequate hydration of the infant during this maneuver is extremely important. The enema may be repeated at 12-hour intervals over several days until all the meconium is evacuated per rectum. If surgical intervention is required because of failure of

Gastrografin enemas to relieve obstruction, resection of the distended terminal ileum is performed, the meconium pellets are flushed from the distal small bowel by N-acetyl cysteine, and ileostomies or a Bishop-Koop anastomosis is performed. The microcolon can then be irrigated with saline or mineral oil to increase its lumen size, and anastomosis or closure of the Bishop-Koop chimney can be deferred for several weeks or months.

Necrotizing Enterocolitis (NEC)

Clinical Manifestations. Necrotizing enterocolitis is a disease predominantly affecting infants who are highly stressed. The most typical infant is the small premature baby with respiratory distress syndrome, though other problems such as sepsis can predispose the infant to the development of NEC. Blood flow is maximized to the brain and cardiopulmonary circulation under conditions of stress, producing a low mesenteric blood flow. This relative ischemia to the gut may be the result of an active process in the infant such as is seen in the diving animals. Mucosal ischemia occurs if the infant is fed, especially if hyperosmolar formula is used. The mucosa is then vulnerable to invasion by gas-producing bacteria. The hallmark of this disease therefore is the development of gas in the intestinal wall, producing the x-ray finding of pneumatosis intestinalis. In severe cases, gas may extend into the portal vein. Ischemia may progress to involve all layers of the bowel wall with gangrene and perforation. Clinically the first sign of the development of NEC is formula intolerance, characterized by residual formula in the stomach just before a feeding. Discontinuing feeding at this stage may interrupt the ischemic process in the bowel wall. Or the infant may show signs of progressive peritonitis with erythema and edema of the abdominal wall, distension, and tenderness, followed by increasing acidosis, sepsis, disseminated intravascular coagulation, and ultimately death. Distended, stationary loops of bowel can be seen on x-ray. Progression to free intraperitoneal air is seen following perforation. X-rays should be repeated every 6 to 8 hours as the infant is followed clinically.

Treatment. Free intraperitoneal air and signs of peritonitis are obvious indications for surgery. Surgery may also be needed for the infant without these signs who is showing progressive deterioration, acidosis, and a falling platelet count in spite of maximum supportive measures of intravenous fluids, antibiotics, and sometimes mechanical ventilation. In equivocal cases, tapping the peritoneal cavity and lavaging with small quantities of saline may aid in the decision to operate if the fluid withdrawn contains white cells and/or bacteria. Resection of frankly gangrenous bowel should be carried out, and in the vast majority of cases, the intestinal ends are brought out as stomas. A decompressing gastrostomy is also performed. "Second-look" procedures may be necessary in 1 to 2 days to assess bowel that is marginally viable at the time of initial exploration.

Total parenteral nutrition is carried out for at least 2 weeks postoperatively, after which oral feedings of small volumes of isosmolar formula are gradually introduced. Full recovery of ischemic mucosa may take many weeks, so that prolonged malabsorption with diarrhea may occur. Strictures may develop as the intestine heals. This is particularly true in the defunctionalized colon. Some of these strictures may resolve spontaneously with time, but evaluation of the bowel with contrast is necessary before reestablishing intestinal continuity.

Intussusception

Intussusception is a common cause of intestinal obstruction in the infant. It is most often observed in babies between 8 and 12 months of age and is slightly more common in males. The most frequent type is probably the result of hypertrophy of the Peyer's patches in the terminal ileum from an antecedent viral infection. The hypertrophied lymphatic patch becomes drawn into the lumen of the terminal ileum and is progressively moved into the ascending and transverse colon. Polyps, malignant tumors, such as lymphoma, and Meckel's diverticulum may act as lead points for intussusception; such intussusceptions are usually confined to the ileum and occur in older infants and children.

Clinical Manifestations. Since intussusception is frequently preceded by a gastrointestinal viral illness, the onset may not be easily discovered. Typically the infant develops paroxysms of crampy abdominal pain and intermittent vomiting. Between attacks he may act completely well, but as symptoms progress, increasing lethargy becomes apparent. Blood mucus ("currant jelly stool") may be passed per rectum. Ultimately if reduction is not accomplished, gangrene of the intussusceptum occurs.

The pathognomonic physical finding is an elongated mass in the right upper quadrant or epigastrium with an absence of bowel in the right lower quadrant (Dance's sign). The mass may be seen on plain abdominal x-ray but is more easily demonstrated on barium enema. Barium outlines the leading portion of the intussusception, giving a "coiled-spring" appearance.

Treatment. The barium enema can also be used for hydrostatic reduction of the intussusception. Signs or peritonitis constitute a contraindication to barium enema reduction. The enema bag is suspended no higher than 1 m above the rectal catheter, and barium is allowed to enter the colon by gravity. Approximately 60 to 70 percent of the time, the barium column will push the intussusception before it and accomplish reduction. Free reflux into multiple small bowel loops and an infant who abruptly becomes well are characteristic of successful reduction. Unless both of these signs are observed, it cannot be assumed that the intussusception is reduced, and preparation for exploration are made.

In some centers glucagon is injected intravenously in an effort to aid reduction by relaxing the smooth muscle of the intestine. A recent multicenter controlled study has not shown any increased success in children who received glucagon.

If hydrostatic reduction is successful, the infant is kept on intravenous fluids for approximately 48 hours before restarting oral fluids. The incidence of recurrent intussusception is low. Failure to reduce the intussusception mandates surgery. Exploration is carried out through a right lower quadrant incision, delivering the intussuscepted mass into the wound. Reduction can usually be accomplished by gentle distal pressure, milking the bowel out of the intussuscipiens, never pulling it out. The blood supply to the appendix is often compromised, and appendectomy is performed. Resection of frankly gangrenous bowel is carried out without attempting reduction of the intussusception. As a rule, primary ileocolic anastomosis can be performed after resection.

Intravenous fluids are continued until peristalsis returns. If resection is necessary, prophylactic antibiotics are also administered for 72 hours.

Duplications, Meckel's Diverticulum, and Mesenteric Cysts

Duplications

Duplications can occur at any level in the gastrointestinal tract but are found most commonly in the ileum. They may be long and tubular, but are usually cystic masses lying within the leaves of the mesentery and sharing a common wall with the intestine. Symptoms may include a palpable mass and/or frank intestinal obstruction. Torsion may produce gangrene and perforation, and subtle or massive bleeding may occur. This bleeding comes from ulceration in the duplication or adjacent intestine if the duplication contains ectopic gastric mucosa.

The ability to make a preoperative diagnosis of duplication usually depends on the presentation. Sonography and technetium pertechnetate scanning are the two most helpful diagnostic tests. For short duplications, resection of the cyst and adjacent intestine with end-to-end anastomosis is easily accomplished. If resection of long duplications would compromise intestinal length, multiple enterotomies and mucosal stripping in the duplicated segment will allow the walls to collapse and become adherent. An alternative method is to divide the common wall using the GIA stapler, forming a common lumen. This should not be done in duplications which contain ectopic gastric mucosa.

Meckel's Diverticulum

A Meckel's diverticulum is the persistence of a portion of the embryonic omphalomesenteric duct. It is located on the antimesenteric border of the ileum, usually a short distance (within 2 feet) from the ileocecal valve. It may be found incidentally at surgery or may present with inflammation masquerading as appendicitis. Like the duplication, ectopic gastric mucosa may produce ileal ulcerations which bleed and lead to the passage of maroon-colored stools. Diagnosis may be made by technetium pertechnetate scans when the patient presents with bleeding. Surgical resection may be by wedge excision of the diverticulum and transverse closure of the ileum if the base is narrow. If wedge excision would compromise the ileal lumen, sleeve excision and end-to-end ileo-ileostomy are performed.

Mesenteric Cysts

Mesenteric cysts are similar to duplications in their location within the mesentery. However, they do not have any mucosa or muscular wall. Chylous cysts may result from congenital lymphatic obstruction. Mesenteric cysts can cause intestinal obstruction or may present as an abdominal mass. Sonography may suggest the diagnosis. Surgical removal is accomplished by resection of the adjacent intestine, partial excision or marsupialization being reserved for cysts involving a large portion of the mesentery.

Hirschsprung's Disease

Hirschsprung's disease is characteristically a disease of the male infant. The defect is the absence of ganglion cells in the rectum and the rectosigmoid. The precursors of the ganglion cells are neural crest cells which migrate into the intestine from cephalad to caudad. The process is completed by the twelfth week of embryonic life, but the migration from midtransverse colon to anus takes 4 weeks. This increases the time period of vulnerability for failure of migration and accounts for the fact that most cases of aganglionosis involve the rectum and rectosigmoid. Longer segments of absent ganglion cells may also occur, and total colonic aganglionosis, although rare, is also seen. Sex incidence is equal in these cases of long-segment Hirschsprung's disease.

Aganglionic colon does not permit normal peristalsis to occur. Functional obstruction therefore supervenes, and the infant may present with complete colon obstruction or with a devastating enterocolitis. The presentation may, however, be much more subtle, with constipation and abdominal distension and sometimes, though not invariably, poor nutrition.

Diagnosis. Infants with Hirschsprung's disease will usually fail to pass meconium in the first 24 hours of life, although this history is often impossible to obtain. Added to this fact that it takes several weeks before the proximal colon containing ganglion cells hypertrophies and dilates enough to show the characteristic change in size of the aganglionic portion of the colon on barium enema. Barium enema is therefore often normal in the newborn infant. In older infants and children, barium enema will show the size contrast between the dilated ganglionic colon and the distal constricted rectal segment. The barium enema in total colonic aganglionosis shows a markedly shortened colon, often in the form of a question mark.

Rectal biopsy makes the definitive diagnosis of Hirschsprung's disease. A suction biopsy technique is available which provides a small piece of mucosa and submucosa.

The diagnosis may also be made by histochemical staining of cholinesterase in the ganglion-cell/nerve complex.

Treatment. Treatment is surgical in all cases. Pull-through procedures have been performed in the newborn period in several centers. Most pediatric surgeons prefer to perform a colostomy in the newborn period and wait for a period of growth (to 10 kg) before doing definitive surgery. In the older infant and child who have been belatedly diagnosed it is important to allow the distended hypertrophied colon to return to a normal size before pulling it through the narrow pelvis. This is usually accomplished by waiting 3 to 6 months after a colostomy is performed.

Three pull-through procedures are currently in use for treating Hirschsprung's disease. The first of these is the original Swenson procedure, in which the aganglionic rectum is carefully dissected in the pelvis and removed down to the anus. The ganglionic colon is then anastomosed to the anus via a perineal approach. Variations of Swenson's technique have been devised by Duhamel and Soave. In the former, dissection outside the rectum is confined to the retrorectal space and the ganglionic colon anastomosed posteriorly just above the anus. Martin has modified this procedure by excising the anterior wall of the ganglionic colon and the posterior wall of the aganglionic rectum, using a stapling device. This avoids the

accumulation of stool in the blind aganglionic rectum. In Soave's operation, dissection is entirely within the rectum. The rectal mucosa is stripped from the muscular sleeve, and the ganglionic colon is brought through this sleeve and later amputated at the anal level. Autoanastomosis is achieved by healing between the colon serosa and the circular muscle of the rectal sleeve. Boley performs a suture anastomosis between the distal colon and the anal mucosa and completes the operation in one stage. These three procedures can also be adapted for total colonic aganglionosis; the ileum is used for the pull-through.

Imperforate Anus

Imperforate anus affects males and females with equal frequency and occurs once in 20.000 live births. Failure of descent of the urorectal septum in embryonic life produce a variety of anorectal and cloacal anomalies. The level to which this septum descends determines the separation of the urinary and hind-gut systems. Broadly classified, the imperforate anus is characterized as "high" or "low" depending on whether the rectum ends above the levator ani muscle complex or partially descends through this muscle. The rectum usually ends in a fistula. In high imperforate anus in males the fistula usually ends in the prostatic urethra. In females the interposed vagina is the site of entry of the fistula. The low lesions have a fistula to the perineum. In males this is seen in the median raphe of the scrotum or penis, and in females the commonest perineal fistula ends at the posterior fourchette. Since the rectum has descended through the levator complex in low lesions, only a perineal operation is required; this situation occurs in 50 percent of males and 90 percent of females. Such children will be expected to be continent since the "muscle of continence", the levator ani muscle, and the rectum are in a normal relationship to each other.

Infants having high imperforate anus require a colostomy in the newborn period, with some kind of pull-through procedure performed after growth to about 10 kg, which renders the technical procedure easier.

Diagnosis of imperforate anus is not difficult. The location of the fistula site may sometimes be a problem. Beading of mucus or meconium along the median raphe of the perineum and scrotum denotes a low imperforate anus in the male. Air in the bladder, voiding of gas or meconium, and retrograde urethrogram with contrast may demonstrate the urinary fistula of the high imperforate anus in the male. In the female, since most of the lesions are low, careful examination of the perineum, fourchette, and vestibule of the vagina will locate the fistula in most cases. A high fistula in the female may terminate in the vagina and will be harder to demonstrate. If a single perineal opening is seen in the female, a cloacal abnormality is present with urethra, vagina, and rectum opening into a common urogenital sinus. The "upside-down films" of Wangensteen and Rice are often confusing and not definitive and now are seldom used. If there is any doubt of the level, it is much safer to perform a colostomy than ruin any chance for continence by an injudicious perineal operation.

A number of pull-through procedures have been developed for high imperforate anus. The hallmark of each of these operations is the location of the levator ani muscle, or so-called puborectalis portion of this muscle, and transposition of the rectum anterior to this muscle. The rectum is sutured to the skin of the perineum in the normal position of the anus. The Kiesewetter-Rehbein procedure utilizes an abdominal approach, with division of the fistula from above. The mucosa is then stripped from the imperforate rectum and the colon pulled

through the mucosal sleeve to the perineum. In the Stephens approach, the dissection proceeds posteriorly through a transsacral approach. The chances of continence from each of these procedures have not proved to be very good, and mucosal anal prolapse has been a particular problem. Recently Pena and DeVries have devised a posterior approach, dividing the levator ani and external sphincter complex in the midline posteriorly and bringing down the rectum after sufficient length is achieved. The muscles are then reconstructed and sutured to the rectum. This procedure is too new to determine whether continence will be better achieved, but prolapse has been less of a problem for these patients, and the procedure shows promise for an improvement in the poor surgical results.

Jaundice

General Considerations

Neonatal jaundice is not itself pathologic. In fact physiologic jaundice, as is implied by the name, affects many infants. This self-limiting condition is characterized by elevation of the indirect, or nonconjugated, fraction of bilirubin. Jaundice with surgical implications usually means elevation of the direct reacting, or conjugated bilirubin, fraction. Mixed hyperbilirubinemia (elevated indirect and direct fractions) results from hepatic parenchymal dysfunction or extrahepatic ductal obstruction. The evaluation of the jaundiced neonate attempts to differentiate between obstructive and nonobstructive causes. Cholestatic syndromes, due to impaired hepatocellular function, may result from a variety of infectious or metabolic causes. Intrauterine viral infections, infections acquired postnatally, and genetically determined disorders such as $alpha_1$ antitrypsin deficiency or arteriohepatic dysplasia (Alagille's syndrome) must all be considered. These heterogeneous conditions, previously grouped as "neonatal hepatitis", result in severe intrahepatic cholestasis. The diagnosis of neonatal hepatitis is nonspecific and has in the past included the entities mentioned above as well as other conditions, certain to be identified, which result in cholestasis and neonatal jaundice. In addition to these causes of jaundice, there are anatomic abnormalities which result in obstruction to the flow of bile from the liver. These include choledochal cyst and biliary atresia. It should be emphasized that although these latter diseases have their principal expression in the extrahepatic ductal system, the liver itself is also involved. Thus, certain forms of neonatal hepatitis, choledochal cyst, and biliary atresia may represent a spectrum of hepatobiliary pathology resulting from a common insult.

Diagnosis. In the laboratory evaluation of the jaundiced infant, it is essential to remember that no single test is absolutely diagnostic. In addition to the routine tests of liver function, laboratory studies must include serologic screening for intrauterine infection, as well as determination of $alpha_1$ antitrypsin activity in the serum. There is overlap in the laboratory results obtained from patients with jaundice resulting from cholestasis and those with ductal obstruction from biliary atresia. Percutaneous needle biopsy is useful in the hands of experienced pathologists, familiar with pediatric hepatic pathology. Giant cell transformation of hepatocytes is seen in both patients with "hepatitis" and those with biliary atresia. In the latter condition, however, there is canalicular bile stasis and proliferation of bile ducts in portal areas, characteristic of obstruction. Radioactive nuclide scanning employing isotopes of ^{99m}Tc has largely replaced ^{131}I rose bengal for biliary imaging. ^{99m}Tc (IDA) scanning has proved to be highly reliable for defining biliary tract patency in those jaundiced infants having syndromes of intrahepatic cholestasis. The appearance of isotope in the gastrointestinal tract

unequivocally establishes patency of the bile ducts. Diagnostic accuracy is enhanced by pretreatment of the infant with phenobarbital. Failure of isotope to appear in the gastrointestinal tract is regarded as an indication for surgical exploration, with the presumption that the diagnosis of biliary atresia will be confirmed at laparotomy. The operative approach to the patient with biliary atresia was pioneered by Kasai, who demonstrated that within the fibrous tissue investing the bile ducts at the port hepatis are found microscopically patent biliary channels which communicate with the intrahepatic ductal system. These patent biliary structures at the porta probably become obliterated at some time around the twelfth week of life, precluding success if surgical correction is delayed beyond this time.

Extrahepatic Biliary Atresia

Treatment. In the vast majority of infants with extrahepatic biliary atresia, the entire extrahepatic biliary tree is affected. A rare patient will have patent hepatic and common hepatic ducts with distal atresia; this is the so-called favorable type which lends itself to establishment of biliary intestinal continuity. However, for 95 percent of patients with biliary atresia, the hepatic ducts are obliterated by fibrous scar where they emerge from the liver. For this group the portoenterostomy procedure offers the only chance for relief of jaundice. It is important to note that biliary atresia does not imply an absence of the extrahepatic biliary system; rather these structures are present as fibrous cords. A successful surgical outcome depends upon transection of the fibrous tissue investing patent biliary channels at the porta. The most reliable anatomic landmark for identification of the fibrous cone of tissue is the bifurcating portal vein. Transection of this tissue outside the liver substance above this vein provides the only opportunity to establish a functional biliary intestinal anastomosis. When the operation is carried out before the twelfth week of life, bile drainage is anticipated.

It has been shown, however, that bile drainage is not equivalent to cure. Hepatic fibrosis may be ongoing even when bile drainage is achieved. Postoperative cholangitis continues to plague infants having successful portoenterostomy procedures. The incidence and severity of cholangitis are reduced by external venting of the surgically created biliary conduit, and by the long-term administration of antibiotics. Despite these measures the problem of ascending infection has not been eliminated.

Cirrhosis and portal hypertension have emerged as new problems in some children relieved of jaundice by functioning biliary conduits. In some, gastrointestinal hemorrhage from esophageal varices has required intervention by endoscopic sclerotherapy or portosystemic shunting.

Based upon the experience reported from centers throughout the world there is little doubt that the infant with conjugated hyperbilirubinemia deserves an expeditious diagnostic evaluation. When the cause of jaundice is unknown or if there is a high suspicion that the underlying cause is biliary atresia, prompt surgical intervention is warranted. Bile drainage is anticipated in the majority of infants operated before the third month of life, and despite remaining difficulties, tangible improvements in survival have been achieved for a condition previously regarded as hopeless.

Biliary Hypoplasia

In some infants, at the time of surgical exploration and operative cholangiography, the extrahepatic bile ducts may appear anatomically normal but have only a minute lumen. This has been termed "biliary hypoplasia" and is associated with a variety of hepatic parenchymal disorders resulting in intrahepatic cholestasis. Included among these are $alpha_1$ antitrypsin deficiency and arteriohepatic dysplasia (Alagille's syndrome). The primary pathology resides within the liver and not in the bile ducts of these patients. The ducts remain small, presumably because of disuse. Portoenterostomy is not applicable for this category of patients.

Choledochal Cyst

Extrahepatic biliary obstruction from choledochal cyst probably results from a localized obliterative process in the distal common duct. This disorder may be a more generalized alteration in the biliary drainage system since many infants develop liver failure despite surgical relief of the obstructed bile flow. The diagnosis is suspected by the presence of an upper abdominal mass associated with jaundice and fever. Ultrasound examination confirms the presence of a cystic mass in the porta hepatis. Rarely, there is also dilatation of the intrahepatic biliary channels. The surgical options include drainage of a choledochal cyst directly to the duodenum or jejunum. However, these procedures are attended by a high incidence of postoperative cholangitis. Further, the choledochal cyst itself has been regarded as a premalignant lesion, and carcinoma arising from the lining of the cyst has been observed. For these reasons surgical excision of the cyst is recommended, with reconstruction by Roux en Y hepatic duct jejunostomy. Cyst excision is facilitated by opening the cyst and excising all except the posterior portion of the cyst wall which lies upon the portal vein and hepatic artery. The inner lining of this portion of the cyst is stripped free and resected. This approach reduces the hazards of the surgical resection since the major vascular structures are not jeopardized.

Deformities of the Abdominal Wall

Embryology

The abdominal wall is formed by four separate embryologic folds, cephalic, caudal, and right and left lateral folds, each of which is composed of somatic and splanchnic layers. Each of the folds develops toward the anterior center portion of the celomic cavity, joining to form a large umbilical ring which surrounds the two umbilical arteries, the vein, and the yolk sac or omphalomesenteric duct. These structures are covered by an outer layer of amnion, and the entire unit comprises the umbilical cord. Between the fifth and tenth weeks of fetal development the intestinal tract undergoes a rapid growth outside of the abdominal cavity within the proximal portion of the umbilical cord. As development is completed, the intestine gradually returns to the abdominal cavity. Contraction of the umbilical ring completes the process of abdominal wall formation. Duhamel has described failure of closure of any segment of the anterior abdominal wall as celosomia. For example, (1) failure of the cephalic fold to close (upper celosomia) results in sternal defects (as congenital absence of the sternum or the pentalogy of Cantrell); (2) failure of the caudal fold to close (lower celosomia) results in exstrophy of the bladder and in more extreme cases, exstrophy of the cloaca. Interruption of central migration of the lateral folds results in omphalocele.

Gastroschisis, originally thought to be a variant of omphalocele, is probably a rupture of intrauterine umbilical hernia.

Umbilical Hernia

Failure of timely closure of the umbilical ring leaves a central defect in the linea alba. The resulting umbilical hernia is covered by normal umbilical skin and subcutaneous tissue, but the fascial defect allows protrusion of abdominal content. Hernias less than a centimeter at the time of birth will usually close spontaneously by one to two years of life. Larger defects may require longer periods of time before spontaneous closure occurs. Some umbilical hernias never close spontaneously. Umbilical hernias are easily recognized as a protrusion of the umbilicus covered by normal skin. Sometimes the hernia is large enough that the protrusion is disfiguring and disturbing to both the child and the family. In such circumstances early repair may be advisable. When the defect is small and spontaneous closure likely, delay of surgical correction until four or five years of age is appropriate. Incarceration is rarely seen in an umbilical hernia. Unlike treatment for inguinal hernia of infants and young children, attempts at reduction of an incarcerated umbilical hernia are unwise. Repair of uncomplicated umbilical hernia is performed through a small curving infraumbilical incision which fits into the skin crease of the umbilicus. The fascial defect is repaired with permanent sutures in the midline. Fascial flaps or other complicated umbilical hernia repairs which have been recommended for adult patients are unnecessary in children. The umbilicus should never be excised in the repair of umbilical hernias in the childhood age.

Patent Urachus

During the development of the coelomic cavity, there is free communication between the urinary bladder and the abdominal wall through the urachus, which exits adjacent the omphalomesenteric duct. Persistence of this tract results in a communication between the bladder and the umbilicus. The first sign of a patent urachus is moisture or obvious urine emission from the umbilicus. Recurrent urinary tract infection may result. The urachus may be partly obliterated, with a remnant remaining beneath the umbilicus in the extraperitoneal position as an isolated cyst. Diagnosis of patent urachus is most reliably made by a cystogram in the lateral projection. Surgical correction is carried out via extraperitoneal exposure of the infraumbilical area. Identification and excision of the urachal tract with closure of the bladder is curative. Urachal cysts are also easily excised from this approach.

Patent Omphalomesenteric Duct

In fetal life, the omphalomesenteric duct is connected through the central wall of the coelomic cavity to the intestinal tract. Normally this duct involutes, but its persistence results in a tubular attachment between the ileum and the umbilicus. Liquid ileal content refluxes through the umbilical defect, soiling the abdominal wall. Diagnosis of a congenital fistula at the umbilicus is made by inspection, probing of the tract, and introduction of radiopaque material into the ostium. Proper surgical treatment consists of elective abdominal exploration with closure of the fistula of the antimesenteric border of the ileum and total excision of the fistulous tract, including its attachment to the undersurface of the umbilicus. Though not an emergency, this procedure should not be postponed, since there is a potential for intestinal volvulus to occur around this intraabdominal structure. Occasionally the peristaltic activity

of the bowel will cause eversion of the intestine through this patent duct. The extruded intestine resembles a small ruptured omphalocele, and the lesion requires careful inspection at the neck of the defect to determine its true nature. In such cases the bowel has, in effect, turned inside out and prolapsed through the patent duct, forming an external intussusception. In this instance, immediate operation with reduction and correction is necessary.

Omphalocele

An omphalocele presents as a mass of bowel and solid viscera in the central abdomen, covered by translucent membrane. The size varies from about 1 cm in diameter to huge defects containing much or all of the abdominal viscera. In these latter, the bowel has lost its right of domain in the abdominal cavity, and temporizing techniques must be used until the abdominal cavity reaches a sufficient size to accommodate the bowel. The diagnosis is made by inspection. Babies born with omphaloceles are prone to other anomalies. Rickham collected reports from 11 large clinics whose combined experience yielded a 67 percent incidence of anomalies. Special syndromes such as exstrophy of the cloaca (vesico-intestinal fissure) and the Beckwith-Wiedemann constellation of anomalies include omphalocele.

Emergency treatment immediately after delivery of an infant with omphalocele consists of covering the lesion with saline-soaked gauze and wrapping the trunk circumferentially. No pressure should be placed on the omphalocele sac in an effort to reduce its contents, because pressure can lead to rupture of the sac or may interfere with abdominal venous return, or impede respiratory effort. In 1957 Grob reported a conservative treatment for omphalocele using mercurochrome, to cause a thick eschar cover. This layer subsequently separate, and epithelization progressed from the periphery of the lesion, gradually covering the omphalocele. Although rarely used today, there are certain indications for this approach using betadine spray. These indications are (1) a newborn with a giant omphalocele and other life-threatening anomalies whose correction takes precedence over repair of the omphalocele; (2) the neonate with other anomalies that complicate a surgical repair of the omphalocele; and (3) newborns with severe associated anomalies that may not be consistent with survival.

In 1948 Gross described a technique whereby the abdominal wall was repaired in two stages. First wide skin flaps were developed which were closed over the intact omphalocele. A second stage was the correction of the ventral hernia at a much later date. Beginning in 1959, Schuster used prosthetic materials sutured to the fascia and covered with skin flaps to bring about a gradual enlargement of the abdominal cavity with staged operative procedures. Subsequently, Allen and Wrenn and Gilbert et al suggested the use of a "silo" of silastic material sutured around the circumference of the defect as an exterior cover uncovered by skin. This technique, refined over the years by many, has proved a basis for temporary coverage of large omphaloceles as well as the exposed bowel in gastroschisis. The main principle in the use of the plastic material is steady pressure with gradual reduction of the plastic envelope over an interval of 5 to 14 days until the abdominal wall closure can be tolerated by the infant. Today most babies, even with large omphaloceles, can be salvaged, and the national survival of about 70 percent reflects the serious anomalies associated with this malformation.

Gastroschisis

Gastroschisis was once thought to be a form of ruptured omphalocele. It was incorrectly described in older literature as a defect of the abdominal wall separated from the umbilicus by a bridge of skin. In fact, the gastroschisis defect which permits escape of the intestines from the abdominal cavity occurs at the junction of the umbilicus and the normal skin. Shaw first suggested that gastroschisis is simply a hernia of the umbilical cord which ruptures after the final formation of the abdominal somatic components. Certainly this thesis is consistent with the findings. Babies with gastroschisis have a large amount of viscera on the surface of an intact abdominal wall that has a small defect at the right edge of the umbilicus which is still held in place by the two arteries and the umbilical vein. The umbilicus has become partly detached, allowing free communication with the abdominal cavity. The intestine lying free outside of the abdominal cavity may be glistening, moist, and normal in appearance, suggesting that the rupture occurred immediately before or during delivery of the infant. More commonly, the intestine is thick, edematous, discolored, and covered with a shaggy exudate; it has been postulated that the intestine escaped from the fetal abdominal confines via the umbilical cord rupture weeks or even months before delivery, and floating freely in the amnion, developed changes in the intestinal wall.

Treatment. All infants born with gastroschisis should be taken to the operating room expeditiously. The intestine can be returned to the abdominal cavity and a secure surgical closure of the abdominal wall achieved primarily in many instances. Some authors report that mechanical stretching of the abdominal wall aids in successful primary closure. For others, particularly those infants whose intestine has become thickened and edematous, the construction of an extraabdominal compartment from silastic sheeting has proved beneficial. As in the surgical correction of omphalocele, this allows gradual enlargement of the abdominal cavity as the intestines are accommodated and the silastic compartment gradually reduced. This latter process takes approximately two weeks and must be carried out carefully to assure that increased abdominal pressure does not cause caval compression which may impede venous return to the heart, nor diaphragmatic pressure preventing normal respiratory excursion. Intestinal function may not occur for several weeks, and is especially delayed in shaggy edematous bowel. In these babies the advent of intravenous alimentation has been life-saving. Although the condition was once uniformly fatal, gastroschisis patients can now usually be saved.

Exstrophy of the Cloaca (Vesicointestinal Fissure)

Exstrophy of the cloaca represents one of the severest forms of embryologic derangement. In infants with cloacal exstrophy, the normal ventral closure of the pelvis and the wall is imperfect. Major components of the cloacal exstrophy are (1) omphalocele, (2) exstrophy of the bladder, (3) external intestinal fistula through the bladder (omphalomesenteric duct), (4) epispadias in the males, (5) imperforate anus, and (6) foreshortened colon. In addition there is often an associated orthopedic deformity of the distal leg and foot. The summation of these physical defects is such that many of the newborns are not hardy enough to survive.

Early surgical intervention in these patients becomes necessary when extensive intestinal prolapse through the fistula causes intestinal obstruction. In such circumstances an ileostomy is required. The colon is temporarily exteriorized as a mucous fistula. The omphalocele is closed primarily or treated with a topical agent which promotes epithelialization. There is no urgency about repairing the exstrophied bladder, and reconstruction of the urinary system need not be completed until the patient is two or three years of age. While most patients suffer a number of physical limitations, many have the potential for functional rehabilitation, which justifies aggressive surgical efforts on their behalf.

Congenital Deficiency of the Abdominal Musculature (Eagle-Barrett Syndrome; Prune-Belly Syndrome)

Congenital deficiency of the abdominal musculature is a rare anomaly occurring in males. In severely affected infants, there is marked wrinkling of the skin of the lower part of the abdomen and little or no muscular substance detectable beneath it. In addition to the absent abdominal muscles, the bladder is large and the ureters are dilated and tortuous. The kidneys may be hypoplastic, but there is usually adequate renal parenchyma.

Advances in techniques of urinary reconstruction have improved the prognosis for this group. Temporary urinary diversion with subsequent stage reconstruction, by shortening, tapering, and reimplantation of the ureters, and reduction in the size of the bladder has been successful in rehabilitating the urinary tract. Various surgical procedures have been devised to tighten the lax abdominal musculature and reduce the redundancy of the abdominal wall, thereby providing adequate support. This approach has changed materially the dismal outlook that most of these children faced a decade ago.

Inguinal Hernia

Inguinal hernia results from a failure of closure of the processus vaginalis, which is normally obliterated in males by two or three months of age. The processus is a fingerlike projection of the peritoneum which accompanies the testicle as it descends into the scrotum. Infants are at particular risk from incarceration of a hernia. The internal inguinal ring is narrow; therefore, intestine finding its way into the hernial sac in the inguinal canal can become trapped within the hernial sac. When there is diagnostic confusion between an incarcerated hernia and a hydrocele, a rectal examination with simultaneous abdominal palpation of the internal inguinal ring will delineate the structures passing through the internal ring into the inguinal canal. Using the vas deferens as a constant reference point, the presence of intestine adjacent to the vas between the examining fingers confirms the diagnosis of a hernia. Most often the hernia can be reduced. The infant is sedated, and moderate, bimanual pressure is applied by compressing the sac from below while a gentle counterforce downward is provided from the examiner's hand above the inguinal ring. Occasionally, these hernias will reduce spontaneously after sedation is given and the continuous struggling and crying are terminated. Surgery should be delayed 48 hours, until the local edema secondary to incarceration has resolved. If the hernia cannot be reduced, or, in cases of obvious intestinal obstruction, emergency operation, with reduction and repair, is necessary.

An inguinal hernia in the female usually is indicated by the appearance of a nontender groin mass. The mass represents an ovary herniated into the patent sac. Although the gonad can usually be reduced into the abdomen by gentle pressure, it often prolapses in and out until surgical repair is carried out. In some patients the ovary and fallopian tube constitute one wall of the hernial sac (sliding hernia), and in these cases the ovary can be effectively reduced only at the time of operation. Because of the frequency of bilaterality, contralateral exploration is recommended for all infants under one year of age except those in whom it is precluded by a coexisting medical condition or in whom the operation on the symptomatic side is unduly difficult.

Hydrocele is often associated with inguinal hernia. The hydrocele may communicate with the peritoneal cavity via the patent processus vaginalis and, therefore, wax and wane in size. Alternatively the hydrocele may be encysted and confined to the scrotum or to the inguinal canal. Usually a simple hydrocele does not require operation unless it is shown after 1 to 2 years of observation that it is not undergoing spontaneous regression.

Genitalia

Cryptorchidism

The term "undescended testicle" describes that testicle which has been interrupted in its normal route of descent into the scrotum. Such a testicle may reside in the posterior abdomen, in the internal inguinal ring, in the inguinal canal, or even at the external ring, but is never in the scrotum. The testicle begins as a thickening on the urogenital ridge in the fifth to sixth week of embryologic life. In the presence of the Y chromosome the testicle stimulates the development of the normal male ducts including the vas deferens, the seminal vesicles, and the prostate gland. The fetal testicle also secretes the Müllerian inhibiting substance which causes regression of the developing female genital tract. The testis is generally formed by the fourth fetal month and begins its migration downward to approach the internal ring at approximately five months' gestation. In the seventh and eight months the testicle descends along the inguinal canal into the upper scrotum, and with its progress the processus vaginalis is formed and pulled along with the migrating testicle. At birth approximately 95 percent of infants have the testicle normally positioned in the scrotum, and by a month of life approximately 99 percent of male infants have a normally positioned testicle. The etiology of the testicle's failure to descend is unknown.

A distinction should be made between the undescended testicle and the ectopic testicle. An ectopic testis by definition is one that has passed through the external ring in the normal pathway and then has come to rest in an abnormal location either overlying the rectus abdominis or external oblique muscle, or the soft tissue of the medial thigh, or behind the scrotum in the perineum. A congenitally absent testicle results from failure of normal development or an intrauterine accident leading to loss of blood supply in the developing testicle.

Clinical Manifestations. The history in patients with undescended testicles may be confusing because of inadequate observation on the part of the parents or the presence of an active retractile testicle. In most patients with a unilateral undescended testicle the testicle can be felt in the inguinal canal or in the upper scrotum. Occasionally, the testicle will be difficult

or impossible to palpate, indicating either an abdominal testicle or congenital absence of the gonad. In some instances, one can palpate the vas deferens entering the scrotum with a small amount of tissue at its end. This may indicate a vascular accident with a poorly formed or absent testicle on that side. In patients with bilateral undescended testicles, it is appropriate to study the serum gonadotropin since the serum luteinizing hormone is elevated in those patients without gonadal tissue. In rare instances an ectopic testicle can be located on the abdominal wall, in the thigh, or in the perineum.

Indications for surgical placement of the testicle in the scrotum are (1) diminished spermatogenesis, (2) malignant degeneration, (3) increased trauma (to a testicle located at the pubic tubercle), (4) increased incidence of torsion, and (5) psychological. Woolley has gathered known information on malignant degeneration of the testicle and has summarized it as follows: (1) the patient with unilateral or bilateral undescended testicle is at increased risk for malignant degeneration of the testicle; (2) surgical or medically induced testicular descent does not protect the individual from testicular malignancy; however, surgical repositioning of the undescended testicle allows easier observation and examination so that malignancy can be promptly detected; (3) the reason for malignant degeneration is not established, but the evidence points to an inherent abnormality of the testicle that predisposes it to incomplete descent and malignancy rather than a malignancy by an abnormal environment; (4) the right testis is more frequently involved in malignant change than the left; (5) the most common tumor in association with undescended tests is seminoma.

It appears that the male with bilateral undescended testicles is infertile. It is also suggested by most authors that the influence of the body temperature is significant in diminishing spermatogenesis. The scrotal position seems to encourage normal spermatogenesis. Mengel and co-workers studied 515 undescended testicles by histology and demonstrated a decreasing presence of spermatogonia after two years of age. Consequently it is not recommended that the undescended testicle be surgically repositioned if it is not properly descended by two years of age.

Treatment. Chorionic Gonadotropin. The use of chorionic gonadotropin has been most effective in patients with bilateral undescended tests, suggesting that these patients are more apt to have a hormone insufficiency than children with unilateral undescended testicle. If there is no testicular descent after a month of endocrine therapy, operative correction should be undertaken.

Orchiopexy. Unilateral undescended testicle should be approached surgically before the second year of life. The dissection in the inguinal canal allows repair of a coexisting inguinal hernia and access to the retroperitoneum so that the vas deferens and spermatic vessels can be carefully dissected. The main principles of operation are (1) to dissect the floor of the inguinal canal, the underlying retroperitoneum, the vas deferens, and the vessels, gaining adequate length for repositioning the testicle; (2) to pass the testicle and spermatic structures beneath the transversalis fascia and the epigastric vessels, thereby bringing the internal ring down behind the external ring, decreasing the distance customarily traversed by the testicle; (3) to create an adequate pouch so that the previously unused scrotum will accept the testicle; (4) to fix the testicle in the scrotum by passing it through the Dartos fascia, allowing it to reside in a compartment created between this fascia and the skin.

When the testicle is located high in the retroperitoneum and the spermatic vessels are very short, staged procedures which gradually stretch the vessels are usually unsatisfactory. As an alternative to removal, Fowler and Stevens proposed dividing the spermatic artery and vein, preserving all collateral blood supply along the vas deferens, and bringing the testicle into the scrotum on a pedicle containing the deferential artery. Others have substantiated this as a viable alternative, and it is to be recommended over orchiectomy or allowing the testicle to remain in the abdomen (with the threat of malignant degeneration). Use of microvascular techniques to transpose the testicle from the abdomen to the inguinal area, though promising, has not yet been adapted to young children.

Some patients who have an absent testis are greatly bothered by this anatomical deficiency. Gel-filled prostheses of all sizes are now available and can be simply inserted into the scrotum achieving normal appearance and a normal structure for palpation.

Any patient who has an undescended testicle corrected surgically should be examined yearly by his surgeon until his midteen years. At that time the individual should undergo thorough explanation about the possibility of malignant degeneration and be instructed in self-examination, which should be carried out at least twice a year for life.

Intersexual Abnormalities

Normal sexual differentiation occurs in the sixth fetal week as a result of genetic factors which influence the development of the gonads. The testis-determining gene is probably located on the Y chromosome, but this is not yet firmly established. In the recent past the HY antigen has been identified as a possible testis-determining factor. In every fetus, Wolffian (male) and Müllerian (female) ducts are present until the onset of sexual differentiation. The development of internal ducts into the male series or the female series is directed entirely by the fetal testis which secretes both testosterone and Müllerian inhibiting substance. Testosterone stimulates maturation of Wolffian duct structures into epididymis, vas deferens, and seminal vesicles; simultaneously, the Müllerian inhibiting substance produces regression of the female structures. In the absence of the fetal testis, the Müllerian system proceeds to full maturation. Any disruption of the orderly steps in sexual differentiation may be reflected clinically as variants of the hermaphrodite syndromes. These may be classified as (1) true hermaphroditism (with ovarian and testicular gonadal tissue), (2) male pseudohermaphroditism (testicles only), (3) female pseudohermaphroditism (ovarian tissue only), and (4) mixed gonadal dysgenesis (usually undeveloped or imperfectly formed gonads) of each sex. Most of these clinical forms present with ambiguous external genitalia, which are usually obvious at birth.

Male pseudohermaphroditism is found in genotypic males with bilateral testes; however, the duct structures of many of these patients differentiate partly as phenotypic males. This group of disorders can result from defects in androsynthesis or incomplete Müllerian regression. Female pseudohermaphroditism is most commonly found in those patients with congenital adrenal hyperplasia. These individuals are unable to synthesize cortisol. This deficiency causes ACTH to stimulate the secretion of excessive quantities of adrenal androgen which masculinizes the developing female.

In the differential diagnosis of patients with intersexual anomalies, the following steps are important: (1) evaluation of genetic background; (2) determination of biochemical factors in serum and urine; (3) assessment of the anatomical structures by physical exam and x-ray studies, and when necessary, by laparotomy and gonadal biopsy.

With refined diagnostic techniques, most infants with intersexual abnormalities can be accurately assessed in the first days of life, obviating errors in gender assignment. Subsequently, certain plastic surgical procedures are required to harmonize the external genitalia with the sex of rearing. Operations to reduce the size of the enlarged clitoris have been developed which spare the sensation and function of the clitoris. Plastic procedures to exteriorize the vagina or separate it from the urethra are necessary in patients born with a urogenital sinus. When the male assignment is appropriate for an infant with an ambiguous genitalia, hypospadius repair will be necessary. When contradictory gonads or ovotestes are present, removal of these structures is required to prevent the possibility of malignant degeneration. For psychological adjustment of some teenage male patients with inadequate or absent gonads, the insertion of a testicular prostheses may prove beneficial. Children with endocrine deficiency may require lifetime exogenous supplementation. Prompt recognition of infants with intersexual anomalies, followed by appropriate sex assignment and proper treatment, avoids the social and psychological derangements which have occurred in the past because of delayed diagnosis or inappropriate gender assignment.

Neoplastic Disease

Approximately 12 percent of deaths in children are attributable to malignant disease. Solid tumors account for just over half, while leukemia is responsible for death in the remaining children. Wilms' tumor and neuroblastoma are the most commonly encountered tumors. The presenting clinical features are similar, with both characterized by upper abdominal masses, some of which assume gigantic proportions. The establishment of the proper diagnosis before surgery is essential because the technique of resection, subsequent management, and prognosis varies according to the type of neoplasm.

Wilms' Tumor

Approximately 500 new cases of Wilms' tumors are recorded in the USA each year. The usual presentation is that of an asymmetric mass in the upper abdomen. The incidence is greatest in the first two years of life. Both sexes are affected equally. While many imaging modalities have proved useful for the assessment of these tumors. the intravenous pyelogram remains the most important diagnostic study. The collecting system typically shows marked distortion, with the calyces appearing to be pulled apart and fragment. Rarely, the tumor invades the renal pelvis and obstructs the ureter, but usually drainage from the kidney is unimpaired. Occasionally, the kidney involved by Wilms' tumor will not function on intravenous pyelogram. The inferior vena cava should be evaluated by venacavagram, ultrasound, or CT scan in those patients in whom renal vein or caval tumor thrombus is suspected. This is especially applicable in patients with large tumors.

Wilms' tumors are no longer regarded as surgical emergencies, but prompt nephrectomy with ipsilateral lymph node dissection is the preferred treatment. A generous transverse incision provides adequate exposure for the removal of the primary tumor, and

allows palpation and observation of the opposite kidney as well. When possible, the renal pedicle is approached before the tumor is manipulated in order that the renal vein be ligated early in the procedure, thus reducing the risk of tumor embolization.

After surgical resection, patients with Wilms' tumors receive chemotherapy and/or radiation therapy, according to the stage of their lesion. Those patients with tumors limited to the kidney, and no external disease, require no radiation. Chemotherapy with actinomycin D and vincristine has changed the outlook remarkably for patients with residual or recurrent disease. Actinomycin D has a synergistic effect when coupled with radiation therapy. While chemotherapy and radiation therapy have contributed to improved survival in children with Wilms' tumor, surgical excision remains the primary treatment. Wilms' tumor is one of the most curable child cancers, with a 5-year survival of approximately 80 percent. For those patients with tumor confined to the kidney or regional lymph nodes (Stages I or II), the cure rate is 95 percent.

Neuroblastoma

Neuroblastoma is the third most frequent malignancy of children, accounting for approximately 11 percent of all childhood cancer. Neuroblastomas arise from the sympathetic nervous system, the typical sites being the adrenal glands and posterior mediastinum. They may also originate from neural tissue in the neck or pelvis. Fifty percent of neuroblastomas occur in patients under one year of age, and eighty-five percent have occurred by age three years. The younger children have a better prognosis. Patients seen before the age of one year have an overall survival rate of approximately 75 percent. This favorable survival is realized even when there is metastatic disease to bone marrow, liver, or skin (Stage IV, S). Osseous metastases are ominous, and survival is rare if there is bony involvement. Evidence is accumulating that immune mechanisms conferred by the mother are responsible for the extraordinary spontaneous regression and maturation seen when neuroblastoma appears in the early months of life.

Abdominal neuroblastoma is characteristically an irregular hard mass which often crosses the midline. An intravenous pyelogram will usually show lateral and inferior displacement of the kidney, but the calyceal distortion characteristic of Wilms' tumor is almost never seen. Thoracic tumors of neural origin are located in the posterior mediastinum, a distinguishing feature which is readily determined on a lateral chest film.

Neuroblastomas lose their ability to store norepinephrine and epinephrine within tumor tissue. Because there is a continuous synthesis and breakdown of these substances, vanilylmandelic acid (VMA) levels are increased. VMA concentration has been a useful biochemical test in both diagnosing the tumor and following the course of treatment. Surgical excision is the treatment for neuroblastoma, regardless of the site of origin. In the abdomen and retroperitoneum this may prove impossible because the tumor often grows around the celiac axis and mesenteric artery. When this pattern of growth is encountered, the bulk of the tumor should be surgically excised (debulking procedure). Most neuroblastomas respond to a variety of chemotherapeutic agents and are sensitive to radiation therapy, but these agents have not altered survival to the extent accomplished with other childhood tumors.

Teratoma

Teratomas are composed of tissue from all three embryonic germ layers although one cell type may predominate. Though teratomas may arise in any part of the body, a predilection for certain sites is well recognized. Those tumors arising in the chest are usually found in the anterior mediastinum and are generally benign. Teratomas arising in the retroperitoneum usually present as a mass in the abdomen or flank. Such tumors may mimic other abdominal malignancies in clinical presentation. The goal of therapy is complete surgical removal. If contiguous spread has not occurred, the cure rate for retroperitoneal teratoma is high

Sacrococcygeal Teratoma

Sacrococcygeal teratoma usually appears as a mass originating from the buttocks and projecting forward to distort the perineum. The mass may vary in size from a few centimeters to a massive tumor, approaching the size and weight of the newborn infant. Myelomeningocele, lipoma, and neural tumors with intraspinal extension should be considered in the differential diagnosis. The majority of these tumors are benign, but the potential for malignant degeneration is high. After the second month of life, the majority will show malignant components. Therefore, complete excision in the early days of life is essential, and can usually be accomplished without damaging the rectum or genital structures. If the tumor is completely excised, the cure rate is excellent, but malignant lesions do not respond to x-ray therapy or chemotherapy.

A form of sacrococcygeal teratoma exists which is not evident externally but resides in the presacral space and projects inwards toward the rectum and pelvis. Discovery of these tumors is often delayed until symptoms of rectal obstruction appear. The late discovery of this variant of sacrococcygeal teratoma allows malignant degeneration, and the cure rate is considerably decreased in this form.

Rhabdomyosarcoma

Once thought to originate only from striated muscle, rhabdomyosarcoma is now recognized as an embryonic tumor which can arise from a variety of mesenchymal tissues. The commonest sites of origin are the head and neck, the extremities, and the genitourinary system. This rapidly growing neoplasm invades surrounding structures. Metastases occur to regional lymph nodes and the lungs. Wide local excision is now recognized as the optimal surgical treatment for localized forms of rhabdomyosarcoma. Radical extirpation in the form of amputation does not improve survival. Tumors arising in the female urogenital tract may present externally through the vagina as a multilobulated mass (sarcoma botryoides). Radical ablative surgery is usually not necessary for these lesions. This change in therapeutic concept has followed the extraordinary success of combined chemotherapy with actinomycin D, vincristine, and Adriamycin for this disease.

Hepatoma

Two main forms of malignant liver disease occur in children, hepatoblastoma and hepatocellular carcinoma. Hepatoblastoma is slightly more frequent and its prognosis is somewhat better than that of hepatic carcinoma. The child usually has an abdominal mass

often associated with anemia, low-grade fever, and anorexia. The patients are rarely jaundiced, and most tests of liver function are normal. Alpha fetoprotein, a substance usually not detectable in the serum after the first few weeks of life, is present in patients with hepatoblastoma and is a useful biological marker. Before surgery, complete radiographic evaluation, including selective hepatic angiography, is essential. Hepatic resection offers the only hope for cure. Recent attempts to reduce the bulk of massive tumors by pretreatment with chemotherapy have met with limited success. The value of radiotherapy is limited. Extended radical hepatic surgery has increased the survival of children with primary liver tumors. For hepatoblastoma approximately 60 percent of the children have a surgically accessible tumor, and of these patients, half are curable, for an overall survival of 30 percent. With respect to hepatocellular carcinoma, the outlook is not as favorable, with only 15 percent being cured by any surgical procedure.

Trauma

The National Safety Council documents that 103.303 individuals died of accidental causes in 1977. Nearly one-tenth of these fatalities, or 9.602 deaths, occurred in children one to fourteen years of age. Accidents accounted for 46 percent of all pediatric deaths in 1977, more than the six other leading pediatric diseases combined - cancer, congenital anomalies, pneumonia, heart disease, homicide, and meningitis. Motor vehicle injury is the most common cause of accidental death (20 percent) in children, while drowning (8 percent), burns (5 percent), and firearms (1 percent) account for a significant segment of the remaining group. Not evident in the mortality statistics is the number of children who sustain injury, are treated, and recover with a subsequent disability. However, the National Safety Council estimates that 220.000 children sustained injuries or died in 1979 as a result of motor vehicle-related accidents alone.

Improved methods of communication, immediate prehospital care, and rapid transportation to regional centers increase probability of survival following a major traumatic injury. Because of the complex requirements for resuscitation and treatment of the injured child, it is essential that each regional referral center establish a systematic approach to the care of the young accident victim. Personnel trained in pediatric trauma are needed to plan, implement, and manage the injured child. Integration of the multidisciplinary human and material resources of each designated hospital is essential.

Management of the pediatric trauma patient requires immediate recognition and treatment of life-threatening injuries to the head, thorax, and abdomen. Recognition that the child with severe head injury manifests a different pathophysiologic response from that of the adult forms the foundation for successful therapy. Aggressive therapy of elevated intracranial pressure results in improved survival; 9 percent of the victims of head trauma die, while 88 percent make a good to excellent recovery. Thoracic trauma occurs in a third to a half of all children sustaining multiple trauma. Blunt abdominal trauma is common in the pediatric population. The spleen and liver are the most frequently injured organs, with an associated mortality of 5 to 12 percent.

Consideration of the unique requirements of the injured child during evaluation and therapy improves outcome and minimizes mortality. Aerophagia proceeding to gastric dilatation can compromise respiration or mimic an abdominal injury; therefore nasogastric

decompression is mandatory in all children with significant injury. The temperature of injured children must be monitored carefully because of the relatively large surface area to body mass ratio. Hypothermia potentiates the deleterious effects of the patient in shock. Hypovolemic shock with cellular hypoperfusion is treated with crystalloid infusion at a rate of 20 mL/kg, IV push. Urine output (1 mL/kg/h) in children should be maintained. Once initial assessment and resuscitation have been accomplished, diagnostic measures with conventional x-rays and CT scan may be important. Prompt movement to a pediatric intensive care unit is essential unless a decision for operation is necessary. Complete rehabilitation of the child and the family should be the ultimate goal of every pediatric trauma center.