

MINISTRY OF HEALTHCARE OF UKRAINE
Ukrainian Medical Stomatological Academy

Educational lectures for recipients of higher education of the 5th course of study

<i>Educational discipline</i>	Surgery, including pediatric surgery
<i>module №2</i>	Pediatric surgery
<i>Theme of the lesson</i>	Purulent diseases of the lungs and pleura, the bone and joints, the soft tissues. Tumors in the children.
<i>Course</i>	5
<i>Faculty</i>	International faculty

POLTAVA - 2021

The number of teaching hours 2

1. Scientific - methodical substantiation topics

The topic is very important for future doctors in their professional activity, positively influences the students in their attitude to the future profession, forms professional skills and experience as well as taking as a principle the knowledge of the subject learnt.

2 Educational lectures aim.

1. To master the list of diseases which cause intrathoracic tension.
2. To recognize the basic clinical manifestation of intrathoracic tension (intrapulmonary and intrapleural).
3. To differentiate intrathoracic tension depending on the reason of its origin.
4. To master the forms of hematogenous osteomyelitis, complications and consequences causing abnormalities of the bone and joint system.
5. To recognize the basic clinical manifestations of septic arthritis (metaphyseal osteomyelitis) for babies for children above 2 and atypical forms.
6. To master the list of purulent diseases of soft tissues characteristic for new-born ones, stages of the development of septic process.
7. To recognize the basic clinical manifestations of purulent diseases of skin, subcutaneous fat tissue and the like.
8. To master diseases which cause a formation of soft tissue.
9. To recognize the basic clinical manifestation of formations of soft tissue.
10. To recognize the basic clinical manifestation of tumours of bones, distinguish the signs of malignant regeneration.
11. To master the characteristic signs of syndrome of "palpable tumour of abdomen" at nephroblastoma and neuroblastoma.
12. To distinguish nephroblastoma and neuroblastoma depending on clinical manifestation and stage of the course of diseases.
13. To recognize the basic clinical manifestations of tumours of mediastinum.

3. Development of personality of future specialist (educational aims), relevant aspects of:

1. In the process of lighting themes to promote further development of analytical and logical thinking in students, to deepen the logical connection between the children's surgery as a discipline and the clinics of Pediatrics and surgery, noting the peculiarities of surgical diseases in pediatric surgery.
2. To educate responsibility for the timeliness and accuracy of medical action.
3. To be able to set psychological contact with the parents of a sick child.
4. Carefully adhere to the principles of medical ethics and deontology.

4. Interdisciplinary integration.

Names of previous disciplines	Obtained skills
1. Anatomy	Describe the anatomy of the abdominal, thoracic cavity and musculoskeletal system. To assess the features of possible variants of the anatomical structure of the thoracic and abdominal cavities, to determine the features of the structure of bones in different age periods.
2. Histology	Know the histological picture of the abdominal, thoracic cavity and musculoskeletal system. Be able to determine the features of the histological picture of different parts of the gastrointestinal tract, the chest and bone in children of different age groups

3. Operative surgery	To depict schematically the peculiarities of surgical interventions in children. To determine the features of topographic anatomy of the thoracic, abdominal cavities, musculoskeletal system in children of different age groups; To justify operative dissections and interventions depending on the pathology and age of the child
4. Propedeutics of childhood diseases	Possess the technique of examining a child with diseases and tumors of the thoracic, abdominal cavity, musculoskeletal system. To assess and demonstrate the knowledge of clinical and laboratory studies, the main symptoms of inflammatory diseases of the thoracic and abdominal organs, the main clinical symptoms are characteristic of diseases of the musculoskeletal system.
5. Patanatomy	Infectious pathologoanatomical changes in diseases and tumors and diseases of the respiratory system, gastrointestinal tract, musculoskeletal system. Identify specific pathological changes, their sequence for tumors of the thoracic, abdominal cavities in children of different ages and tumors of the musculoskeletal system.
6. Biochemistry	Demonstrate laboratory methods of examining a child with diseases and tumors of the abdominal, thoracic cavity and musculoskeletal system. To evaluate the data of clinical and biochemical analyzes: glucose in blood tests, urine; Protein in the blood serum, urine; Microelements
7. Pathological physiology	Describe pathological changes in diseases and tumors of diseases of the respiratory system, gastrointestinal tract, musculoskeletal system. To determine the main points of etiology, pathogenesis in acute surgical diseases, tumors of the thoracic, abdominal cavities in children of different ages.

5. Plan and organization. Structure lectures

The main stages of lectures and their contents.	Type lecture. Means revitalization students. Materials methodological support.	Time
1. The preparatory stage. Determining the relevance of the theme: educational purposes lectures and motivation		5%
2. The main stage		
Submission of lecture material on the plan 1. The forms of acute destructive pneumonia 2. The complications belong to the pulmonary form of acute destructive pneumonia. 3. the clinical forms of acute gematogenic osteomyelitis. 4. the most frequent symptom of metaepiphyseal acute gematogenic osteomyelitis.	Overview summarizing the lecture.	85%-90%

5. The main reasons of origin of purulent-septic diseases in new-born ones. 6. The methods of conservative and operative treatment of hemangioma and lymphangioma.. 7. Surgical and combined treatment of malignant tumours of soft tissue. 8. Clinical manifestations and treatment of neuroblastoma at children. 9. Clinical manifestations and treatment of neuroblastoma at children. 10. Clinic and diagnostics of tumours of the mediastinum.		
3. Concluding Stage Summary Lectures, Sharing conclusions.	Training literature. Tasks questions.	5%

6. The content of the lecture material.

PULMONARY ABSCESS

Abscesses of the lung occur when pulmonary parenchyma becomes obstructed, infected, and then suppurative and necrotic. Abscesses may be single or multiple, and be caused by a single organism, usually aerobic, or by anaerobic flora, usually mixed. *Klebsiella* and staphylococcal pneumonias often result in multiple abscesses, which occur infrequently in pneumococcal, streptococcal and *H. influenzae* pneumonias. Multiple abscesses may also be associated with tuberculous or mycotic infections. More often multiple abscesses occur in patients with such chronic pulmonary disease as cystic fibrosis or bronchiectasis, or with illnesses associated with diminished host resistance (agammaglobulinemia, agranulocytosis, chronic granulomatous disease of childhood, leukemia).

Solitary lung abscesses may be tuberculous, may follow pneumococcal or staphylococcal pneumonia, may stem from infected cysts or may be found in sequestered pulmonary tissue. Most commonly, however, a solitary lung abscess follows aspiration of a foreign body or other infected material or such surgical manipulations as tonsillectomy, adenoidectomy and tooth extractions. Abscesses associated with aspiration of tissue or foreign bodies are usually infected by bacteria normally found in the nasopharynx, such as anaerobic bacteroides, spirochetes, and various streptococci, generally not group A.

Whatever the cause, the pathologic evolution of abscess formation is similar. Initial inflammatory changes are followed by suppuration and thrombosis of the local blood vessels, which result in necrosis and liquefaction. Granulation tissue forms around the periphery of the abscess and may succeed in walling off the area, but more commonly the abscess will rupture into a bronchus and be evacuated. Contents of the abscess may be coughed up, or aspirated into other parts of the pulmonary tree, with additional abscess formation. Sputum is usually fetid, may separate into layers, and usually contains elastic fibers.

Peripheral abscesses may involve the adjacent pleura, with development of a plastic or occasionally a serofibrinous pleurisy. Abscesses may rupture into the pleural cavity and produce empyema.

On occasion, pulmonary abscesses may occur within interlobar fissures, where they are usually well encapsulated and respond poorly to antimicrobial therapy.

CLINICAL MANIFESTATIONS. The onset of lung abscess is occasionally insidious, but more commonly there is the sudden appearance of fever, cough and chest pain, often associated with dyspnea and tachypnea. The fever curve is often septic in type, and leukocytosis is usually marked. Physical examination may or may not reveal an area of pulmonary consolidation, depending on the location of the

abscess and its size. At an early stage roentgenographic examination will usually show a wedge-shaped area of consolidation.

In untreated patients the abscess will often rupture into a bronchus within a week to 10 days after onset, with production of purulent or putrid sputum; hemoptysis is common in older children. At this time roentgenographic examination will usually reveal a cavity, with or without a fluid level, surrounded by an area of consolidation. Spontaneous drainage of the abscess may result in disappearance of symptoms within about a month. During this interval, clubbing of the fingers may appear and recurrent hemoptysis may be seen.

TREATMENT. Adequate treatment of pneumococcal pneumonia with penicillin will usually prevent pulmonary cavitation. With staphylococcal and klebsiella pneumonias, cavitation often occurs despite treatment but rarely requires special therapy. It is generally enough if the underlying pneumonia is treated vigorously with suitable antimicrobial therapy. When a foreign body is suspected, bronchoscopic examination should be performed promptly for verification and removal, if possible. Bronchoscopy should be done also, as soon as an abscess ruptures into a bronchus, to aspirate the purulent material and to secure bacteriologic cultures by aerobic and anaerobic techniques. Repeated bronchoscopic aspirations may be needed for the patient who continues to cough up large quantities of purulent material. Intensive and appropriate antimicrobial therapy should be continued for at least 2 weeks. The instillation of proteolytic enzymes or antibiotics into the abscess cavity has not contributed significantly to therapy. As long as the patient continues to bring up sputum, he should receive postural drainage and physical therapy to the chest.

When patients do not respond to initial bronchoscopic aspiration and intensive antimicrobial therapy, repeated aspirations of the abscess may lead to eventual closure of the cavity. If conservative management has not given satisfactory results in 1 month, surgical removal of the affected segment or lobe is usually carried out.

PULMONARY GANGRENE

Gangrene of the lung is extremely rare in children. It occasionally follows measles, and is seen in persons with severe immunologic deficits. The onset is usually sudden and is associated with early pulmonary hemorrhage; there is rapid development of pneumothorax and putrid empyema, death occurring quickly. Treatment consists of adequate pleural drainage and intensive antimicrobial therapy.

PLEURISY

Inflammatory processes in the pleura are usually divided into three general types: dry or plastic, serofibrinous or serosanguineous, and purulent pleurisy or empyema.

SEROFIBRINOUS PLEURISY

Serofibrinous pleurisy is most commonly associated with infections of the lung or with inflammatory conditions of the abdomen or mediastinum. Less commonly it is found with such mesenchymal diseases as lupus erythematosus, periarteritis or rheumatic fever. On occasion this type of effusion is seen with neoplasms of the lung, pleura or mediastinum, which may be primary or metastatic; tumors are, however, more commonly associated with a hemorrhagic pleurisy. Of infectious diseases, tuberculosis has been the most frequent cause of serofibrinous effusion, but in population groups where mycobacterial disease occurs infrequently, pneumococci have become the most common infectious agents.

CLINICAL MANIFESTATIONS. Since serofibrinous pleurisy is often preceded by the plastic type, the early signs and symptoms may be those of the latter illness. As fluid accumulates, pleuritic pain may disappear and the patient become asymptomatic so long as the effusion remains small, or there may be only the signs and symptoms of the underlying disease. If a large amount of fluid collects, there may be cough, dyspnea, tachypnea, orthopnea or cyanosis. Physical findings depend to some degree on the amount of effusion. Dullness to flatness may be found on percussion. There is a decrease or absence of breath sounds, a diminution in tactile fremitus, a shift of the mediastinum away from the affected side, and, on occasion, fullness of the intercostal spaces. If the fluid is not loculated, these signs may shift with changes in position. In infants, physical signs are less definite; sometimes, instead of decreased or absent breath sounds, bronchial breathing will be heard. If extensive pneumonia is present, rales and rhonchi may also be audible. Friction rubs are usually present only during the early or late plastic stage. The process is usually unilateral.

Roentgenographic examination shows a more or less homogeneous density obliterating the normal markings of the underlying lung. Small effusions may cause only obliteration of the costophrenic or

cardiophrenic angles or a widening of the interlobar septa. Examination should be performed both in the supine and in the upright positions to demonstrate a shift of the effusion with change in position. The decubitus position may also be helpful.

DIFFERENTIAL DIAGNOSIS. Thoracentesis should always be done when pleural fluid is known to be present or is suspected. Examination of the fluid is essential to identify acute bacterial infections and may disclose tubercle bacilli. Furthermore, thoracentesis can differentiate between serofibrinous pleurisy, empyema, hydrothorax, hemothorax and chylothorax. In hydrothorax the fluid has a low specific gravity, below 1.015, and only a few mesothelial cells rather than leukocytes. Chylothorax and hemothorax usually have fluid distinctive in appearance. It is not possible to differentiate serofibrinous from purulent pleurisy without bacterial examination of the fluid. The fluid of serofibrinous pleurisy is clear or slightly cloudy and contains relatively few white cells and, occasionally, some red cells. Serofibrinous fluid may rapidly become purulent; its nature may depend on the time during the course of illness when thoracentesis is performed.

COURSE. Unless the fluid becomes purulent, it usually disappears relatively rapidly, particularly with bacterial pneumonias. It persists somewhat longer with mesenchymal diseases and tuberculosis and may remain or recur for a long time with neoplasms. As the effusion is absorbed, adhesions usually develop between the two layers of the pleura, but no functional impairment results. Pleural thickening may develop and is occasionally mistaken for small quantities of fluid or for pulmonary infiltrates. Residual pleural thickening may persist for a long time. In general, however, the process disappears, leaving no residua.

TREATMENT. The treatment is that of the underlying disease. When a diagnostic thoracentesis is done, as much fluid as possible should be removed. If the underlying disease is adequately treated, there is usually no necessity for further drainage, but if sufficient fluid reaccumulates to embarrass the patient's respiration, repeated drainage should be performed.

PURULENT PLEURISY

(*Empyema*)

Purulent pleurisy, or empyema, is an accumulation of pus in the pleural spaces. At present the condition is most often associated with pneumonia due to staphylococci, less frequently with pneumococci and *H. influenzae*. In pediatric practice, empyema is most frequently encountered during infancy.

The disease may be produced also when a lung abscess ruptures into the pleural space, by contamination introduced from trauma or thoracic surgery, or rarely by mediastinitis or by the extension of intra-abdominal abscesses.

PATHOLOGY. Most commonly, purulent pleurisy is an extensive process, consisting of a series of loculated areas involving a large portion of one or both pleural cavities. Thickening of the parietal pleura occurs. If the pus is not drained, it may dissect through the chest wall (*empyema necessitatis*), into lung parenchyma, producing bronchopleural fistulas and pyopneumothorax, or into the abdominal cavity. Pockets of loculated pus may eventually develop into thick-walled abscess cavities, or, as the exudate organizes, the lung may collapse and be surrounded by a thick, inelastic envelope.

CLINICAL MANIFESTATIONS. Since most purulent pleurisy occurs early in the course of bacterial pneumonia, the initial signs and symptoms are primarily those of the underlying disease. Patients treated inadequately or with inappropriate antimicrobial agents may have an interval of a few days between the clinical phase of pneumonia and the evidence of empyema. In infants, manifestations of the disease may consist only of moderate exacerbation of respiratory distress. The older child is apt to appear more toxic and in greater respiratory difficulty. Physical and radiologic findings are identical to those described for serofibrinous pleurisy; the two conditions can be differentiated only by thoracentesis, which should always be performed when empyema is suspected. The maximum amount of pus obtainable should be withdrawn. The physical appearance of pus produced by different organisms is not particularly distinctive; cultures must always be obtained and Gram-stained smears examined for the presence of microorganisms. Staphylococci are usually numerous and thus easily identified; pneumococci and *H. influenzae* occasionally are present only in small numbers, particularly if antimicrobial therapy has been given previously.

COMPLICATIONS. With staphylococcal infections, bronchopleural fistulas and pyopneumothorax commonly develop. Other local complications encountered with any bacterial agent include purulent pericarditis, pulmonary abscesses, peritonitis secondary to rupture through the diaphragm, osteomyelitis

of the ribs, and such septic complications as meningitis, arthritis and osteomyelitis. With staphylococcal empyema, septicemia occurs infrequently; it is often encountered in *H. influenzae* and pneumococcal infections.

TREATMENT. If pus is obtained by thoracentesis, closed drainage should be instituted immediately and controlled either by an underwater seal or by continuous suction. A catheter with the largest possible internal diameter should be inserted into the site where accumulation of pus is suspected; sometimes several tubes are required to drain loculated areas. Closed drainage is usually necessary only for a week or so, even though small amounts of material will continue to drain after this time; this material is usually formed in response to the presence of the tube in the pleural cavity. There is no need to withdraw the tube gradually; rather, it should be removed all at once.

The introduction of fibrinolytic agents or proteolytic enzymes commonly produces severe systemic reactions in small children, and they do not appear to promote drainage substantially. If the chest tube is of sufficient caliber and is kept clear, a free flow of pus is obtained. The instillation of antibiotics into the pleural cavity does not improve results obtained with systemic antimicrobial therapy alone and is associated with local reactions. No attempt should be made to control empyema by multiple aspirations of the pleural cavity rather than by closed continuous drainage.

Systemic antimicrobial therapy is required; the selection of the antibiotic should be based on the in vitro sensitivities of the responsible organism. Staphylococcal empyema in infancy is best treated by parenteral routes with methicillin or, when applicable, with penicillin G. Pneumococcal infection responds to penicillin, and *H. influenzae* to ampicillin. There is no advantage in the use of multiple antimicrobial agents. With staphylococcal infections, resolution of the process is slow, and systemic antimicrobial therapy is required for 3 or 4 weeks. In patients with inadequately treated empyema, extensive fibrinous changes may take place over the surface of the collapsed lungs; these may require decortication at a future date. If pneumatoceles form, no attempt should be made to treat them surgically, or by aspiration, unless they reach sufficient size to embarrass respiration or become secondarily infected.

PNEUMOTHORAX

Pneumothorax in the neonatal period may be related to factors incident to birth and be associated with interstitial emphysema and pneumomediastinum. In staphylococcal pneumonia in infancy the incidence of pneumothorax is relatively high. Aside from the accidental introduction of air into the pleural cavity during thoracentesis, pneumothorax is uncommon during childhood. Pneumothorax may occur in pneumonia, usually in connection with empyema; it may also be secondary to pulmonary abscess, gangrene, infarct, rupture of a cyst or an emphysematous bleb (as in asthma), foreign bodies in the lung and external thoracic trauma or surgical procedures. In association with mediastinal emphysema it is an occasional complication of tracheotomy.

Pneumothorax may be associated with a serous effusion (*hydropneumothorax*) or a purulent effusion (*pyopneumothorax*). In pneumothorax the lung collapses toward the hilus, unless prevented by adhesions. Bilateral pneumothorax is rare.

CLINICAL MANIFESTATIONS. The onset is usually abrupt. When the pneumothorax is extensive, there may be pain, dyspnea and cyanosis. In infancy both symptoms and physical signs may be difficult to recognize. If the pneumothorax is only moderate in extent, there may be little displacement of intrathoracic organs and few or no symptoms.

The percussion note over the involved area is tympanitic; on auscultation respiratory sounds are feeble or absent. Larynx, trachea and heart may be shifted toward the unaffected side. The breath sounds may have an amphoric quality if there is an open fistula from air-bearing tissues into the pleural cavity. When fluid is present, there is usually a sharply delimited area of tympany above a level of flatness to percussion. It is important to determine whether the pneumothorax is an open (*tension pneumothorax*) or a closed one. The presence of amphoric breathing or of gurgling sounds synchronous with respirations when fluid is present in the pleural cavity is suggestive of an open fistula. Confirmatory evidence is provided when the pneumothorax fills rapidly after aspiration of it. Another means for determining whether a fistula is open is examination of the aspirated air for its oxygen content. If a fistula is present, the oxygen content of the air in pneumothorax remains constant. If there is no connection with the bronchial tree, the oxygen content is low, since it is rapidly absorbed. The diagnosis can usually be established by roentgenographic examination).

DIFFERENTIAL DIAGNOSIS. Pneumothorax must be differentiated from localized or generalized emphysema, from an extensive emphysematous bleb, from large pulmonary cavities or other cystic formations, from diaphragmatic hernia and from gaseous distention of the stomach. In most instances a simple roentgenogram will be all that is necessary for the differentiation. In the case of diaphragmatic hernia, however, a small amount of barium may be necessary to demonstrate that a portion of the gastrointestinal tract is in the thoracic cavity.

PROGNOSIS AND TREATMENT. The prognosis depends upon the cause. When there is no fistula connecting the air-bearing tissue and the pneumothorax, the air is usually absorbed within a week or so, and no treatment is necessary unless there are symptoms of excessive pressure, in which case the air should be aspirated.

Tension pneumothorax with a communicating fistula is usually best managed with a closed thoracotomy and drainage of the applied air through a catheter whose external opening is kept in a dependent position under water. If the broncho-pleural fistula is large, negative pressure in the drainage tube may be necessary. If the tension pneumothorax is not relieved by this means, surgical closure of the fistula should be considered. Treatment of a coexisting empyema is of course essential.

OSTEOMYELITIS

This disease occurs most often between 5 and 14 years of age and twice as frequently in boys as in girls. In infants under 2 years of age acute hematogenous osteomyelitis differs in many respects from that in older children.

ETIOLOGY AND PREDISPOSING FACTORS. The causative organism in the majority of instances is the hemolytic *Staphylococcus aureus*, though most of the other pathogenic bacteria may also be responsible. Primary lesions are often demonstrable and include furunculosis, impetigo, infected chickenpox and burns, and vaccinations.

PATHOLOGY. Osteomyelitis begins as a hematogenous abscess in the metaphysis, and then, if interrupted, the abscess ruptures subperiosteally and spreads along the shaft of the bone under the periosteum. The infection then penetrates to the bone marrow. The deep layer of the periosteum forms a shell of new bone around the infected shaft. The pieces of dead bone are known as sequestra, and the new bone formed in the periosteum as the involucrum. Sinuses may form between the sequestra and the skin surface, the hip the metaphyseal abscess ruptures into the joint and creates a suppurative arthritis.

CLINICAL MANIFESTATIONS. The onset is usually abrupt, with fever, malaise, and pain with a localized tenderness in the bone at the metaphysis. Shortly thereafter swelling and redness over the affected bone may be present. These signs appear earlier in infants than in older children. The patient is toxic and extremely weak and irritable.

When osteomyelitis follows an infection which has been treated with an antibacterial agent, the clinical course may be modified sufficiently so that the true nature of the lesion may not be suspected until it is well advanced. In addition, inadequate antibacterial therapy of an acute osteomyelitic infection may temporarily abolish the clinical manifestations, but permit the infection to continue in a suppressed state only to become evident days or weeks later.

DIAGNOSIS. There is a leukocytosis of 15,000 to 25,000 cells or more, and the blood culture is usually positive. Roentgenographic examination does not reveal the process for at least 5 days in small children; in older children this period may be as long as 8 to 10 days. At this time there is rarefaction of the involved area, and soon there is evidence of the formation of involucrum.

DIFFERENTIAL DIAGNOSIS. Rheumatic fever, leukemia, primary or metastatic neoplasm, sprain, cellulitis, erysipelas and scurvy are likely to require differentiation. The presence of great toxicity and localized pain suggests osteomyelitis. Usually this is enough to distinguish the condition from rheumatic fever, but a history of involvement of other joints is indicative of the latter disease, as is the response to salicylates. Scurvy produces painful and tender swelling along the shaft of the bone, but roentgenograms of the long bones should be diagnostic. **PROGNOSIS.** The mortality rate from acute pyogenic infections of the bones has decreased since the availability of specific antibacterial agents. The rate is lower in newborn infants than in older infants and children, as is the incidence of chronic and metastatic lesions. Both the course and prognosis depend on early institution of appropriate therapy and continuance of it for an adequate time.

TREATMENT. Like acute pyogenic arthritis osteomyelitis should be handled as a medical and orthopedic emergency. As soon as one or two specimens have been obtained for blood culture, intrave-

nous antibiotic therapy is initiated. In children under 3 years of age either penicillin-resistant staphylococci or gram-negative organisms are likely to be found, so that therapy should be initiated both with a penicillinase-resistant agent such as methicillin or nafcillin or clindamycin and with intravenous therapy is continued until acute systemic manifestations of infection have subsided; oral therapy is thereafter maintained in full doses for 3 to 4 weeks.

Local treatment of choice for early osteomyelitis is immediate surgical drainage of the metaphysis, though some clinicians illogically prefer to wait 24 to 48 hours to evaluate the response to antibiotic therapy. When the abscess has ruptured into the subperiosteal space, chronic osteomyelitis is the inevitable sequel. Watching and waiting in such a situation is attended with considerable risk.

ACUTE INFECTIOUS ARTHRITIS

This condition is most common in the first 6 months of life. It is usually preceded by an infection elsewhere in the body, often in the upper respiratory tract. The causative organism is usually one of the common pyogens, such as the staphylococcus, streptococcus, pneumococcus, or *H. influenzae* and, less commonly, the gonococcus, meningococcus, typhoid bacillus or one of the salmonella group of organisms. The shoulder, hip and other large joints are most commonly affected, but any joint may be involved. Pyogenic arthritis will result in rapid destruction of cartilage and ankylosis of the joint if diagnosis and treatment are delayed.

CLINICAL MANIFESTATIONS. The onset is sudden, with systemic symptoms of sepsis. Local swelling appears rapidly, with muscular rigidity and intense pain on motion of the joint, and, if untreated, is followed quickly by suppuration. When the hip is affected, it may become dislocated with astonishing rapidity.

DIFFERENTIAL DIAGNOSIS. Acute suppurative arthritis must be differentiated from *acute osteomyelitis*. In acute suppurative arthritis even slight motion of the joint is painful, whereas in osteomyelitis the joint may be moved without pain if done carefully. In suppurative arthritis there is ring tenderness around the joint; in osteomyelitis the tenderness is localized to the metaphysis. In the hip the differentiation cannot be made. The roentgenogram may be of no value in early diagnosis. *Rheumatic fever* rarely occurs in infancy and often involves more than one joint; a prompt response to salicylate therapy is suggestive of rheumatic fever. When an acute pyogenic infection of a joint is suspected, the joint should be aspirated and any material obtained cultured. A blood culture should also be obtained.

TREATMENT. The principle of treatment is immediate drainage of the joint. Emergency drainage can be obtained initially by paracentesis of the joint, but when, by smear or culture, the diagnosis of suppurative arthritis is established, prompt surgical drainage of the joint should be done. Appropriate antibiotic therapy is essential.

Mastitis neonatorum.

Engorgement of the breasts is physiologic in newborn infants. Infection may be abetted by undue manipulation of the breasts and is manifest by redness, local heat, swelling and pain. Fever and other general symptoms may also be present. The prognosis is favorable unless septicemia develops. Prophylaxis consists in avoidance of manipulation or other trauma of the engorged breasts. Treatment includes systemic antibiotic therapy and hot compresses applied locally. If an abscess develops, it should be incised and drained. Scar formation after infection may distort the nipple and impair the secreting power of the mammary gland in a female in later life.

Omphalitis.

Inflammation in the umbilical region, which may be caused by any of the pyogenic bacteria, is especially serious because of the danger of hematogenous spread or extension to the liver or peritoneum. The general manifestations may be minimal even when septicemia or hepatitis has resulted. Prevention of infection depends upon maintenance of a clean umbilical field. Daily baths or daily application of triple dye to the umbilical stump and surrounding skin may reduce the incidence of umbilical infection. *Treatment* includes prompt antibacterial therapy and, if there is abscess formation, surgical incision and drainage.

Perianal Abscess (paraproctitis)

Perianal abscess is a not so rare condition seen almost exclusively in infants less than two years of age. Most cases are seen in males' infants. The infant presents with a history of increasing irritability, fever, erythema and induration of the perianal skin. In a period of 48 to 72 hours the area becomes fluctuant. Oral antibiotics are ineffective in controlling the infectious process. It is theorized that a perianal abscess arises from a developmental anomaly in the deep crypts of Morgagni which trap bacterias initiating a cryptitis that proceed to a perianal abscess. This abscess may open or not to become later a fistula in ano. Gut derived organisms are isolated from most cases of perianal abscess. Most abscesses are located laterally equally divided between right and left. Perianal abscesses in children are best treated by incision, drainage and systemic antibiotics. A proportion of patients with perianal abscess later develop a fistula in ano. This fact has led some researchers to propose that primary treatment of perianal abscess in childhood involve a careful search for a coexisting fistula and treatment of this by fistulotomy. Long term recurrence is very rare. Recurrence should prompt a search for associated disorders such as Crohn's, immunodeficiency and autoimmune neutropenia.

TUMORS OF VASCULAR ORIGIN

These are the most common tumors of early life. Most of them are probably hamartomas rather than true neoplasms, and most are benign. They may occur in any organ, but there are sites of predilection: the skin and subcutaneous tissue, skeletal muscle, liver, salivary gland, larynx and bone.

Cavernous hemangiomas are poorly circumscribed, blue or purple elevated tumors which tend to extend more deeply into the subcutaneous tissues than do the capillary hemangiomas. They consist of numerous cystic vascular spaces containing blood. Mixed forms of cavernous and capillary hemangiomas are common; in such lesions regression of the cavernous elements may be slower and less constant than that of the superficial capillary elements. Cavernous elements which do not regress spontaneously are probably best treated by surgical excision. Cavernous hemangiomas may occur in sites other than the skin, e.g., in bone, liver and the tongue. In skeletal muscles they manifest themselves as a diffuse mass, sometimes accompanied by pain; the mass decreases in size with elevation of the part. Foci of calcification may be demonstrable by roentgenographic examination.

A giant hemangioma, usually of the skin or subcutaneous tissue, but infrequently a visceral one, may be accompanied by thrombocytopenia, bleeding and a consumptive coagulopathy. This is most often seen in the first year of life with a large, rapidly growing cavernous hemangioma, but may occur at a later date in children with multiple hemangiomas that do not involute. Treatment, if necessary, has included the administration of corticosteroids, platelet transfusions, replacement of clotting factors, the use of anticoagulants and/or irradiation. Diffuse hemangiomas of an extremity may be associated with hypertrophy of the part and of the associated bone.

Lymphangioma circumscriptum is a rare condition characterized by small groups of vesicles composed of dilated lymphatics in the superficial dermis. It may be present at birth or begin in infancy or childhood.

Diffuse lymphangiomas are poorly circumscribed tumors occurring in the skin, mucous membranes or muscles. They are usually congenital. In the tongue and lips they are responsible for macroglossia and macrocheilia, respectively. Diffuse lymphangioma of an extremity is responsible for one form of elephantiasis (elephantiasis lymphangiectatica). It may involve an entire extremity or only a portion, e.g., the fingers or foot; there may be associated hypertrophy of the bone as well as of the soft tissues of the length as well as the girth of the extremity. Histologically the dilated lymphatic channels may be obscured by abundant fibrous tissue. The more localized lesions should be treated by surgical excision, but recurrences are frequent. Treatment of the more diffuse lesions is apt to be unsatisfactory.

Cystic lymphangiomas (cystic hygromas) are most frequently encountered in the neck (hygroma colli) and in the axillae, but may also occur in the inguinal and retroperitoneal regions. In the cervical region they may extend into the mediastinum, and rarely mediastinal hygromas may occur in the absence of a cervical component. Mesenteric cysts are simply cystic lymphangiomas of the mesentery, and many omental cysts are lymphangiomas. Sacral hygromas may simulate lipomas or sacrococcygeal teratomas; they are sometimes connected with the spinal canal. Enlargement of cystic lymphangiomas may occur by enlargement of the individual cysts, by formation of new cysts or by hemorrhage into the cysts.

TUMORS OF THE SOFT TISSUES

Neoplasms arising from muscle, fat and connective tissue comprise a miscellaneous but important group of tumors of early life. They may arise at almost any site and vary from benign neoplasms such as the lipoma to highly malignant sarcomas; the latter may be so undifferentiated as to preclude accurate

determination of the cell of origin. They may occur at any age. The most frequent manifestation is a visible or palpable mass. Clinical differentiation of benign and malignant neoplasms is often impossible. Every solid mass should be considered malignant until proved otherwise by histologic examination of the excised mass. **Lymphosarcoma** is the most common malignant neoplasm of the gastrointestinal tract in early life. The tumor usually arises in the small intestine, especially the ileum; it may originate in the colon, appendix or even the stomach. The presenting complaint is usually crampy abdominal pain, often accompanied by vomiting and a palpable mass; the mass may be the neoplasm or an intussusception. Morphologically a segment of the bowel may be diffusely infiltrated by neoplastic cells with resultant thickening of the wall and superficial ulceration of lymph nodes should be removed for diagnostic purposes. Removal of the pancreatic tumor is usually unsuccessful because of its multifocal nature. If the diagnosis of an islet cell tumor can be established histologically either from a primary or a metastatic site in a person with this syndrome, total gastrectomy is probably the treatment of choice, even in children. This may result in disappearance of metastatic lesions.

MALIGNANT TUMORS OF BONE

By far the most frequent primary malignant tumors of bone are the osteosarcoma and Ewing's tumor, the majority of which occur between 10 and 25 years of age; males are affected more frequently than females. Osteosarcoma characteristically involves the metaphyseal end of a long bone, whereas Ewing's tumor involves the shaft, but roentgenographic differentiation of these neoplasms is not always possible. Of greater importance, however, is the fact that many, if not all, of the roentgenographic features of these tumors may be duplicated by non-neoplastic lesions of bone. Accordingly, treatment of lesions suspected of being malignant should not be instituted until an unequivocal diagnosis is established by histologic study of the tumor. Moreover, the pathologist is limited in his ability to establish a diagnosis on the basis of histologic studies alone. For example, an actively growing callus about a fracture may closely simulate the histologic appearance of an osteosarcoma, yet correlation of the material obtained at biopsy with the roentgenographic findings may clearly indicate the true non-neoplastic nature of the process. **Thus, the pathologist must evaluate all pertinent clinical, roentgenographic and surgical data before he arrives at a diagnosis.**

Osteosarcoma (osteogenic sarcoma) is more common than Ewing's tumor. It usually begins at the lower end of the femur or the upper end of the tibia or humerus, but may arise at other sites. The presenting complaint is commonly that of pain and swelling of the affected part, which the patient may attribute to trauma.

Roentgenographic studies reveal varying degrees of destruction of bone and of new bone formation. Codman's triangle is a radiopacity at the end of the tumor where the periosteum has been elevated. Neither this finding nor the perpendicular striations of new bone in the subperiosteal neoplasm ("sunray appearance") are always present, nor are they pathognomonic of an osteosarcoma. The level of serum alkaline phosphatase may be elevated.

The neoplasm occupies the medullary cavity and penetrates the cortex to the subperiosteal zone; penetration of the periosteum into adjoining soft tissues may also occur. Histologically the appearance is varied, but consists essentially of atypical mesenchymal cells with varying degrees of formation of collagen, typical or atypical osteoid tissue and true bone. Cartilaginous areas and areas of myxomatous tissue may be present. Osteosarcoma commonly metastasizes to the lungs, although other organs may also be involved; osseous metastases are rare. Amputation appears to offer the best possibility of cure, but the case fatality rate is high. In one series an exceptional 5-year survival rate of 19 per cent has been recorded. Recently the use of one or more chemotherapeutic agents has given encouraging results in the control of metastases and probably should be utilized postoperatively in all patients; such therapy can be administered only in a hospital equipped with facilities to control the adverse side effects of the treatment.

Ewing's tumor may involve the same bones as does osteosarcoma; in addition, there is relatively frequent involvement of the flat bones and the ribs. The initial complaints are often similar to those associated with an osteosarcoma; fever and leukocytosis may occur with either tumor, but are more likely to be associated with Ewing's tumor.

Roentgenographically there is a mottled area of rarefaction, often associated with increased density and periosteal formation of new bone. The latter may be deposited in layers, resulting in an "onion-skin" appearance, but this finding is often absent and may appear in association with other osseous lesions. The

roentgenographic appearance may closely simulate that of osteomyelitis, osteosarcoma, eosinophilic granuloma of bone or metastatic neuroblastoma.

Gross examination of an affected bone usually reveals more extensive neoplastic involvement than was demonstrable roentgenographically. Histologically the tumor consists of sheets of uniform round or oval nuclei with little or no cytoplasm. The neoplastic cells do not form new bone. Extensive areas of hemorrhage and necrosis are commonly present. The histologic appearance may simulate that of a malignant lymphoma or a metastatic neuroblastoma, and every attempt should be made to exclude the presence of an extraosseous primary lesion.

Ewing's tumor usually involves a single bone when first recognized, but ultimately many bones may be affected. Metastases to the lungs are also common. Treatment currently consists in supervoltage irradiation and chemotherapy rather than amputation. The 5-year survival rate should be in the vicinity of 25 per cent or possibly more; occasionally apparent cures have been effected after the development of metastases.

TUMORS OF THE KIDNEY

Wilms' tumor (nephroblastoma) is one of the most common abdominal neoplasms of early life; approximately two thirds appear before the age of 4 years. Although authentic Wilms' tumors do occur in the neonatal period, almost all the tumors reported as such at this early age have been leio-myomatous hamartomas of the kidney; either of these occurring at birth may be accompanied by polyhydramnios. Bilateral renal involvement is uncommon; when it occurs it is usually detectable at the time of the initial diagnosis; otherwise, the second tumor usually appears within a year or so. Wilms' tumor occurs with increased frequency in children with aniridia, hemihypertrophy and Beckwith-Wiedemann syndrome and probably in those with fused kidneys. The incidence of Wilms' tumor with bilateral aniridia has been reported as high as 1:73 as contrasted with the usual rate of 1:50,000 to 1:100,000 and conversely, Wilms' tumors have occurred in as many as 7 of 28 children under 4 years of age hospitalized with aniridia. The aniridia accompanying Wilms' tumor is almost always of the sporadic type and is apt to be associated with other congenital defects, e.g., cataracts, mental retardation and genitourinary anomalies, including cryptorchidism.

The presenting complaint is usually that of an abdominal mass. Abdominal pain and fever may be the first manifestation. Hematuria is relatively infrequent and, contrary to earlier opinions, is probably not a poor prognostic sign. Physical examination reveals a firm, nontender mass in the renal area which may extend down into the iliac fossa but usually does not cross the midline. Hypertension, the pathogenesis of which is not always clear, may be present.

Roentgen examination reveals a soft tissue density which is apt to displace the intestine toward the opposite side. Calcification is infrequent; when present it is apt to be dense and curvilinear, in contrast to the stippled appearance common in a neuroblastoma. Pyelography, preferably including an inferior vena cavagram, usually reveals distortion of the renal pelvis; displacement or extension of the tumor into the inferior vena cava is sometimes apparent. In some instances the kidney on the affected side cannot be visualized by intravenous pyelography. The pyelographic findings are not pathognomonic of a Wilms' tumor but instead indicate the presence of an intrarenal mass. Probably the most important reason for obtaining pyelo-grams is the demonstration of the presence and apparent normality of the opposite kidney.

Macroscopically the tumor usually presents as a bulky circumscribed mass replacing much of the affected kidney and covered externally by the thin renal capsule. With continued growth, however, there may be invasion of the renal pelvis and/or veins and extension beyond the capsule into the perirenal fat, adrenal, diaphragm or colon. On section the tumor bulges beyond the surface of the adjoining kidney. It is yellowish gray and soft, friable or semiliquid as a result of multiple areas of necrosis; it often contains myxomatous areas and foci of hemorrhage. Areas of necrosis may be responsible for cysts containing clear or hemorrhagic fluid. The renal pelvis is usually elongated and distorted, and occasionally masses of neoplastic tissue extend into its lumen. Careful search should be made for islands of neoplastic tissue in the remainder of the affected kidney since there is some evidence to indicate that such multicentric or metastatic lesions affect the prognosis adversely. Metastases occur principally in lymph nodes, lung and liver.

The histologic pattern is variable. There are often broad sheets or cords of undifferentiated mesenchymal cells within which are scattered epithelial-lined tubules; rarely the latter, if incompletely differentiated,

may resemble the pseudopods of a neuroblastoma. Abortive glomeruli are sometimes present. Bands of loose, more differentiated mesenchymal tissue tend to divide the more cellular areas into coarse lobules, and within these bands smooth and skeletal muscle or, less frequently, bone or cartilage may be present. There is some evidence to suggest that those patients in whom the neoplasm is predominantly epithelial have a better prognosis.

The basic treatment is prompt (but not emergency) radical nephrectomy performed under optimal conditions, usually within 24 to 48 hours after discovery of the mass. Undue palpation of the tumor should be avoided. Preoperatively, roentgen films of the chest should be obtained and the presence and apparent normality of the opposite kidney established by pyelography. Compatible blood should be available if needed for transfusion during operation. At the time of surgery, which should be by the transabdominal approach, the contralateral kidney should be carefully inspected and palpated, the abdominal viscera investigated for evidence of neoplasm and extent of the tumor marked by radiopaque clips.

An attempt to determine optimum therapy is currently being conducted by the National Wilms' Tumor Study. The patients are grouped as follows:

Group I: The tumor is limited to the kidney and is completely resected.

Group II: The tumor extends beyond the kidney but is completely resected; e.g., there has been penetration beyond the capsule, involvement of peri-aortic lymph nodes or infiltration of renal vessels outside the kidney, but there is no apparent tumor beyond the margins of resection.

Group III: There is residual nonhematogenous tumor confined to the abdomen. This includes those patients in whom biopsies have been taken of the tumor, in whom the tumor has ruptured before or during surgery, in whom there are local peritoneal implants, nodal involvement beyond the periaortic chain, or in whom the tumor cannot be completely resected.

Group IV: There are hematogenous metastases, e.g., to the lungs, liver, bone or brain.

Group V: There is bilateral renal involvement either apparent at operation or appearing subsequently.

The treatment of all patients entered into the study is randomized. All patients in Group I are treated by excision and the administration of actinomycin D and vincristine; irradiation is no longer routinely used. All patients in Groups II and III are treated by excision, irradiation, actinomycin D and vincristine. Randomization is currently employed in an attempt to determine the optimum duration of chemotherapy which might be utilized in order to effect cure and to avoid any sequelae.

The results of this collaborative study are not yet available, but the improvement in prognosis following excision, irradiation and chemotherapy has been striking. In one series apparent cures were obtained in 47 of 53 patients who had no demonstrable metastases on admission and in 18 of 31 of those with metastases. With rare exceptions, if a child with a Wilms' tumor is alive and well with no evidence of recurrence or metastatic disease two years after removal of the tumor, a cure has been effected. Such patients should be followed indefinitely, however, for the possible development of subsequent neoplasms secondary to the effects of irradiation.

TUMORS OF THE ADRENAL

Neuroblastoma is one of the most common malignant neoplasms in infants and children. Although more than half of them arise from the adrenal or from the retroperitoneal sympathetic chain, the neoplasm may originate at any site along the sympathetic chain, e.g., in the posterior mediastinum, pelvis or cervical sympathetic ganglia; the tumor may also arise from other derivatives of neural crest origin, such as the dorsal root ganglia. Neuroblastoma is primarily a disease of early life; one fourth of the affected persons have their initial manifestations during the first year of life and three fourths before the age of 5 years. It is the most common malignant neoplasm to be identified at birth, and metastases may even be present at this time.

The presenting manifestation of a neuroblastoma arising in the adrenal or in the neighboring sympathetic ganglia is usually an abdominal mass. It often crosses the midline, in contrast to Wilms' tumor. Roentgen examination reveals a soft tissue mass which displaces the kidney on the affected side downward and laterally; focal areas of calcification are often present. Intravenous pyelography characteristically reveals displacement rather than distortion of the renal pelvis; occasionally, as with Wilms' tumor, the pelvis is not visible on the affected side.

Intrathoracic neuroblastomas are almost always located in the posterior portion of the mediastinum at any level and may be responsible for cough, dyspnea and pain in the chest. A mass may be responsible for

separation of the posterior portions of the ribs and some narrowing and erosion of them. In a number of instances, owing to extradural extensions of the mass, there are manifestations referable to compression of the spinal cord. **Pelvic neuroblastomas** usually produce a demonstrable mass which in some instances simulates a sacrococcygeal teratoma. They may be responsible for urinary or rectal obstruction. **Cervical neuroblastomas** usually do not reach a large size before being recognized. They are apt to present as a hard, lobulated mass involving the posterior triangle of the neck or extending both anterior and posterior to the sternocleidomastoid muscle. In some instances fine stippled areas of calcification within the mass are demonstrable roentgenographically.

The majority of patients with neuroblastoma have metastases when the tumor is first recognized. In some instances the presenting complaint is referable to metastases rather than to the primary tumor, e.g., massive hepatomegaly, especially in young infants, or cervical or axillary lymphadenopathy. Persistent pain and fever may occur with osseous metastases even in the absence of roentgenographic changes and may simulate rheumatic fever or rheumatoid arthritis. Involvement of retrobulbar soft tissues, probably secondary to osseous metastases, may be responsible for proptosis and/or ecchymosis of the upper eyelids. One or more bluish subcutaneous nodules may precede other complaints, especially during the neonatal period. Infrequently, only the metastatic disease is apparent during life, no primary site being identified.

The majority of patients with neuroblastoma have elevated levels of catecholamines or of one or more of their derivatives in the urine. There is some evidence to suggest that this may not occur with tumors which arise in the dorsal root ganglia. Relatively few patients with neuroblastoma, however, initially present with signs of functional endocrine activity such as flushing, perspiration, tachycardia and headache. Intractable diarrhea is an uncommon manifestation. Measurements of the urinary excretion of catecholamines and of their metabolites, e.g., 3-methoxy-4 hydroxymandelic acid (VMA) and homovanillic acid (HVA) may be of diagnostic significance and may also be helpful in the demonstration of residual, recurrent or metastatic disease. Cystathioninuria may also be a diagnostic aid but is less reliable.

Uncommonly acute cerebellar encephalopathy may precede, follow or occur concomitantly with the discovery of a neuroblastoma. There are ataxia, weakness of the extremities and oculogyric crises unaccompanied by pleocytosis in the cerebrospinal fluid and with little or no fever. The cerebellar signs may disappear following removal of the tumor, but mental retardation may persist. Although the mechanism responsible for this association is not clear, patients with acute cerebellar encephalopathy should probably be investigated for the possible presence of an inapparent neuroblastoma.

Metastases of neuroblastoma occur by way of the lymphatic and blood streams; regional and distant lymph nodes, the skeletal system and the liver are the most frequent sites of metastatic spread. Pulmonary metastases occur in only about 10 per cent of patients. Osseous metastases are often bilateral; a unilateral lesion may lead to an erroneous diagnosis of a primary neoplasm of the bone. The roentgenographic changes in the skeleton are characterized by areas of destruction and proliferation of new bone, which may closely simulate the appearance of Ewing's tumor, of eosinophilic granuloma of bone or of skeletal involvement in leukemia. There may be extensive mottling of the cranial bones and separation of the sutures, owing to increased intracranial pressure from metastatic invasion of the dura mater. Neoplastic cells are frequently demonstrable in smears of the bone marrow, even in the absence of roentgenographic changes in the bones themselves.

The neuroblastoma is initially an encapsulated neoplasm, but it soon infiltrates adjoining tissues and, if arising in the adrenal or neighboring sympathetic ganglia, may surround the aorta, inferior vena cava, ureter or renal pedicle and render complete surgical removal impossible. Areas of hemorrhage and necrosis are commonly present, as are minute flecks of calcium. Histologically there may be varying degrees of differentiation toward mature ganglion cells or, less frequently, toward chromaffin cells. The least differentiated neoplasms may be misinterpreted as lymphosarcomas, but additional sections of the same tumor will usually reveal better differentiated areas of neoplastic cells embedded in a haphazard manner within a delicate fibrillary tissue or arranged as pseudorosettes. Less frequently, immature or mature ganglion cells or even chromaffin cells may be present.

The prognosis for the child with a neuroblastoma is dependent upon a number of factors, e.g., extension of the neoplasm across the midline and the extent of maturation of the tumor as determined histologically. The poorer prognosis related to abdominal tumors as contrasted with cervical or mediastinal ones is

probably largely dependent upon the more advanced stage of the disease at the time of diagnosis. Age appears to be the single most important factor with respect to prognosis. The over-all survival rate is in the range of 30 per cent and increases to about 70 per cent in those under 2 years of age. Infants less than 1 year of age, even with metastases to the liver, skin and/or bone marrow but without roentgenographic changes in the bones, have good chances of survival. Cures in the presence of demonstrable osseous metastases have been observed infrequently, even in children over 2 years of age.

Complete surgical removal of the primary tumor is the treatment of choice, but even incomplete removal may be followed by a cure; irradiation is probably indicated if all the tumor is not removed. Chemotherapy, especially with vincristine sulfate and cyclophosphamide (Cytosan) may cause striking regression of the tumor and relief of symptoms in children with widespread disease, but its role in the treatment of those with more localized disease has yet to be established. Certainly the known adverse effects of irradiation and the possibly still unknown effects of chemotherapy must be considered in evaluating the optimum therapy for any patient with this unpredictable neoplasm, in which even spontaneous cures may take place.

Sacroccygeal teratomas arise from the region of the coccyx or lowermost part of the sacrum. They are probably derived from the primordial, to-tipotential cells of the primitive knot (Hensen's node) which, during embryonic life, finally comes to rest in the region of the coccyx. These tumors are three or four times more frequent in girls than in boys, and there is a significant increase in the incidence of twinning in families of persons with sacroccygeal teratomas. At least three fourths of the tumors are apparent at birth, usually presenting as a mass at the tip of the coccyx extending externally in the midline or into one or both buttocks. Large tumors, which may exceed the size of the infant's head, displace the coccyx posteriorly and the anus anteriorly. Occasionally the mass is responsible for urinary or intestinal obstruction, but, in contrast to large myelomeningoceles, they are not responsible for neurologic defects in the extremities.

Rectal examination usually discloses a readily palpable mass posterior to the rectum, which is sometimes encircled by it. Roentgenographic examination usually reveals a soft tissue density in the pelvis, sometimes with displacement of the coccyx posteriorly. Areas of calcification or actual bone are demonstrable in about half of the tumors. In contrast to sacral chordomas, roentgenographic evidence of destruction of the sacrum is rare; when present, it is indicative of a malignant neoplasm. Occasionally spina bifida or lumbosacral anomalies are also present. The differential diagnosis includes meningocele or meningomyelocele; pressure on such a sac will cause the fontanel to bulge, or, if this is closed, crying or straining should increase tension within the mass. Neurogenic tumors, e.g., neuroblastoma and ganglioneuroma, may be clinically indistinguishable from a sacroccygeal teratoma. Chordomas are rare in children; they are responsible for destruction of the sacrum and only rarely extend into the buttock. Papillary ependymoma (below) may present as a mass in the sacroccygeal region. Cystic lymphangiomas and hemangiomas may simulate sacroccygeal teratoma, as may duplication of the hind gut. Occasionally a sacro-coccygeal teratoma presents as a red, inflamed mass or as a draining sinus and thus simulates an infected pilonidal sinus.

The neoplasms are connected to the lowermost part of the sacrum or to the coccyx; the coccyx should always be removed with them. Rarely the tumor extends into the vertebral canal. The tumors are usually well circumscribed solid masses containing multiple cystic structures. Histologically they contain a vast array of tissues; fat, neural elements, smooth and skeletal muscle, bone, cartilage and intestinal and bronchial elements are the most frequent. Teratomatous elements such as pancreatic islets or adrenocortical tissue are sometimes present and may, rarely, produce functional manifestations. Most sacroccygeal teratomas are benign, and cures are sometimes obtained even after one or more recurrences. Most of the tumors discovered before 2 months of age consist only of mature or less often of immature fetal elements and are associated with an excellent prognosis, whereas those appearing later usually contain embryonal carcinomatous areas and are highly malignant. Tumors present at birth but not excised until after 4 months of age are more apt to be malignant than those removed earlier. Symptoms of bowel or bladder dysfunction increase the probability that the neoplasm is malignant. Tumors detected after 5 years of age, as in adults, may be benign or malignant. Recurrences or metastases, if they occur, usually do so within two years after operation.

TUMORS OF THE MEDIASTINUM

Mediastinal masses in infants and children are relatively common. If malignant lymphomas are excluded, most of which are accompanied by manifestations in addition to those referable to the mediastinum, approximately three fourths of the mediastinal masses are neurogenic or teratoma-tous neoplasms or non-neoplastic cysts, e.g., duplications of the esophagus and neurenteric or bronchogenic cysts. Approximately 25 per cent of mediastinal masses in infants and children are malignant as compared with 15 per cent of those in adults. Tumors arising in the anterior mediastinum are predominantly teratomas, whereas most of those originating in the posterior mediastinum are neurogenic neoplasms; masses confined to the mid-mediastinum are usually lymphomas or nonneoplastic cysts.

Approximately two thirds of the infants and children with a mediastinal mass are symptomatic; in the others the lesion is a chance finding on a roentgenogram of the chest. Cough, dyspnea, stridor and pain are the most frequent manifestations and are especially apt to occur with teratomas, malignant neoplasms of any type and with nonneoplastic cysts; vascular tumors and benign neurogenic ones are often unassociated with respiratory symptoms.

Mediastinal teratomas are located in the anterior mediastinum, usually in its superior aspect. Many of them apparently arise in the thymus. Rarely they arise within the pericardial sac and simulate a congenital cardiac lesion. Many are benign cystic neoplasms, commonly referred to as **dermoid cysts**, which are cystic variants of the more solid teratomas. Symptoms may not be apparent until adult life. Dyspnea, cyanosis and cough may be manifestations, and expectoration of hair and sebaceous material may occur if the tumor perforates into a bronchus. Infection of the cystic mass may produce symptoms simulating a pneumonic process. Rarely the neoplasm extends into the suprasternal or supraclavicular area. Compression of the superior vena cava causes dilation of the veins of the head, neck and upper part of the thorax. Roentgenograph[^] examination reveals a circumscribed mass extending from the anterior mediastinum into one hemithorax; when teeth or skeletal elements are demonstrable roentgenographically, the nature of the mass is established.

The teratomas may be composed of one or more cysts; less frequently they are predominantly solid tumors. The cysts contain sebaceous material, hair or mucoid material. Histologically almost any type of tissue may be present, especially in the solid neoplasms. Malignant teratomas, nearly all of which occur in males, are usually solid or finely cystic tumors containing actively proliferating, poorly differentiated tissue in addition to more mature elements; metastatic lesions may resemble the primary tumor or consist only of embryonal carcinoma. Mediastinal teratomas should be surgically removed.

Thymomas are rare in children. In adults they sometimes accompany or precede the development of myasthenia gravis. This association is extremely rare in children, as is the association of thymoma with aregenerative anemia. Thymomas may be asymptomatic and discovered only roentgenographically, or they may be responsible for vague retrosternal pain, cough, dyspnea or signs of compression of the superior vena cava. They are usually encapsulated and composed of an admixture of lymphoid and epithelial cells; typical Hassall's corpuscles are rare. True thymomas are usually benign; occasionally they infiltrate and implant on the pleura.

Lymphosarcoma is the most common malignant neoplasm of the gastrointestinal tract in early life. The tumor usually arises in the small intestine, especially the ileum; it may originate in the colon, appendix or even the stomach. The presenting complaint is usually crampy abdominal pain, often accompanied by vomiting and a palpable mass; the mass may be the neoplasm or an intussusception. Morphologically a segment of the bowel may be diffusely infiltrated by neoplastic cells with resultant thickening of the wall and superficial ulceration of lymph nodes should be removed for diagnostic purposes. Removal of the pancreatic tumor is usually unsuccessful because of its multifocal nature. If the diagnosis of an islet cell tumor can be established histologically either from a primary or a metastatic site in a person with this syndrome, total gastrectomy is probably the treatment of choice, even in children. This may result in disappearance of metastatic lesions.

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