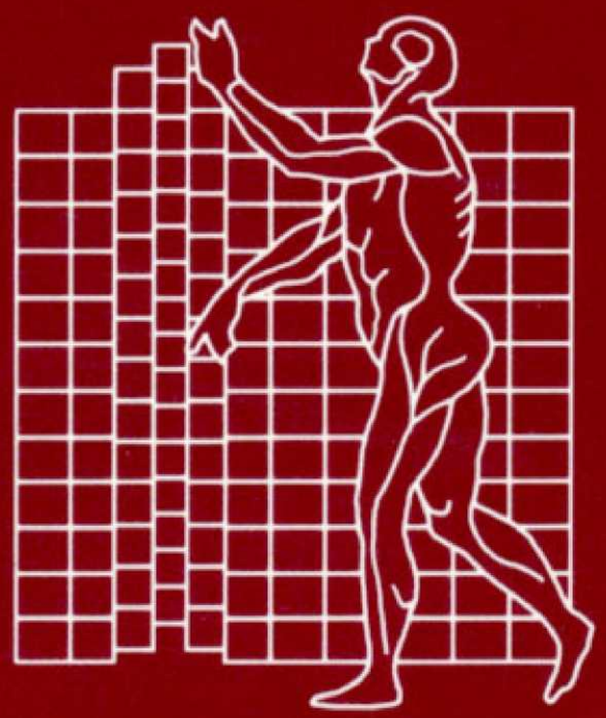
INTRODUCTION TO

CLINICAL MEDICINE



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Introduction to Clinical Medicine

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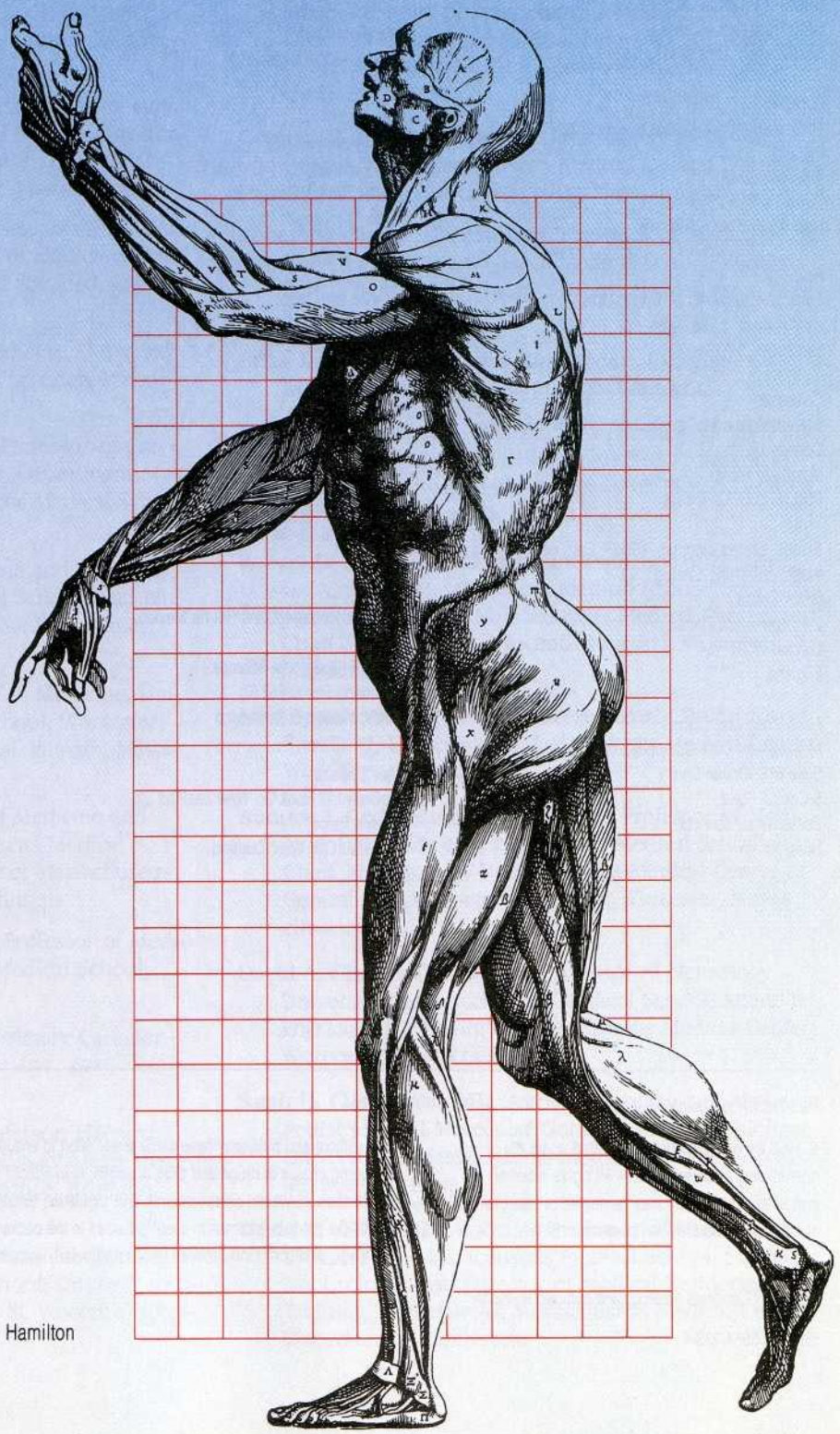

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Clubbing

Earle B. Weiss, MD

Definition

Acquired or hereditary distal clubbing (Hippocratic fingers) is a physical sign characterized by a usually painless, bulbous enlargement of the distal digit of the fingers and at times the toes, accompanied by a softening of the nail bed. Pathologically, the major components of simple clubbing are a fibrous tissue hyperplasia in the segment between the nail and phalanx with lymphocytic extravasation, increased vascularity, and edema. The result is a decrease in the normal obtuse angle between the proximal soft tissue and the nail itself. This phenomenon, although relatively innocuous in itself, is important because its presence may signify a serious underlying disease, and its regression or progression may have important prognostic implications.

History

Typically, the early symptoms and signs of acquired clubbing are overlooked because of their subtle, insidious onset. Hence, digital deformation is generally observed by the physician and not the patient. In rare instances of acute clubbing, as seen with lung cancer or suppurative diseases, unusual tenderness or frank bulbous digital deformity may alert the patient to seek medical advice. Concurrent disease may exist even in severe form without clubbing or hypertrophic pulmonary osteoarthropathy (see later in this chapter); occasionally, acropachy (clubbing) may precede the disorder.

Examination and Measurement

The usual clinical method of diagnosing clubbing, aside from a softening and sponginess of the nail bed, is to measure the angle,

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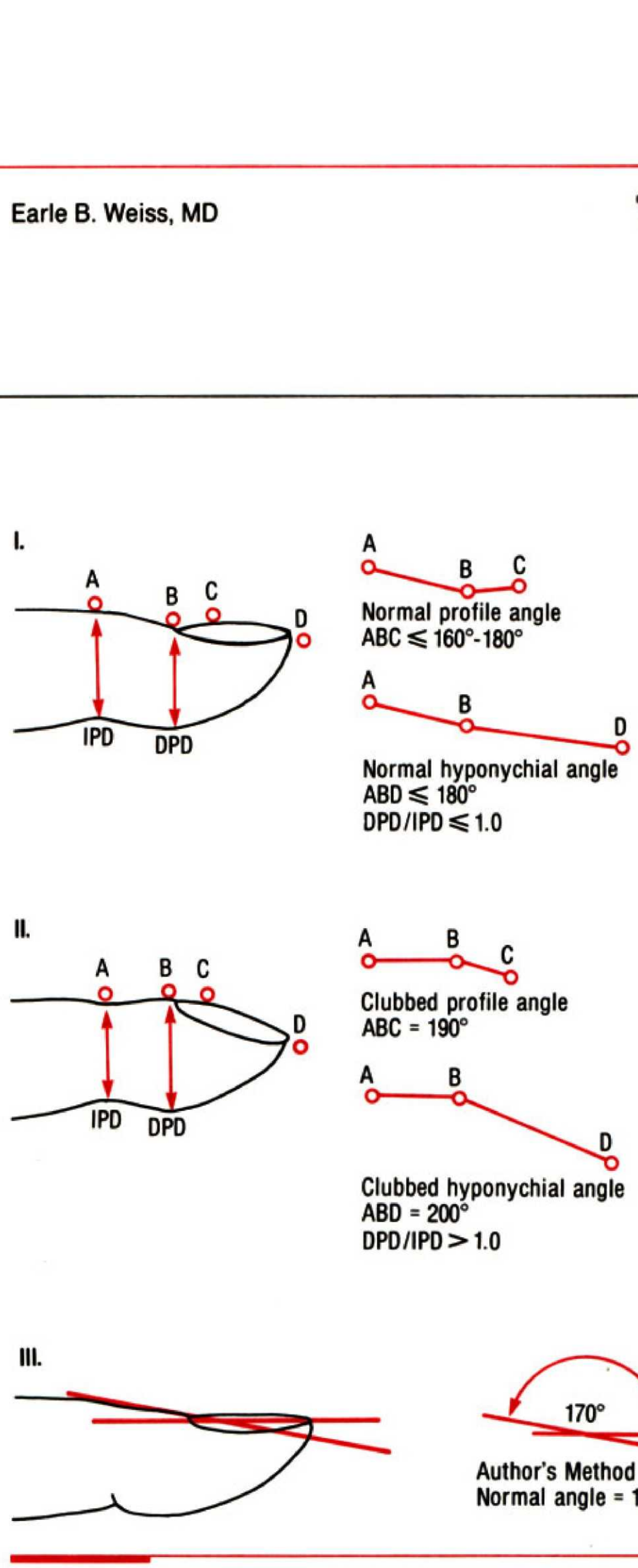

Figure 116-1 Measurement of clubbing. DPD = distal phalangeal depth, IPD = interphalangeal depth.

viewed laterally, described by the base of the nail and the adjacent dorsal surface of the terminal phalanx, the so-called base or profile angle (Fig. 116-1). Coexistent contiguous erythema, warmth, bulk of the terminal tuft, or increased nail curvature is not a sufficient criterion in itself. Normally, this angle is cited to be approximately 160 degrees, although no study has ever validated normal variations by this usual method of visual inspection. Angles greater than 180 degrees are generally considered abnormal. Nonetheless, visual estimates of this angle are largely subjective and therefore unreliable.

Using the following simple bedside method, however, it is possible to obtain accurate and reproducible measurements of the base angle. An affected digit is positioned firmly with its side on a piece of paper. A sharp-tipped pencil is then placed at the base of the finger and, while continuous pressure is applied along the finger tissue, the entire digit is outlined and the nail-proximal digit tissue edge noted. The resulting outline is then easily transcribed into two planes of nail and distal tissue base, from which the base angle can be directly measured. Normal profile angle values observed are similar to those obtained with other techniques: 172.7 degrees ± 4.1 degrees.

Several other features of the clubbing process may be noteworthy. Whereas advanced clubbing generally involves all fingers and toes, the early evolution begins in the thumb and index finger. Other associated physical examination findings include an increase in the nail curvature in coronal and longitudinal

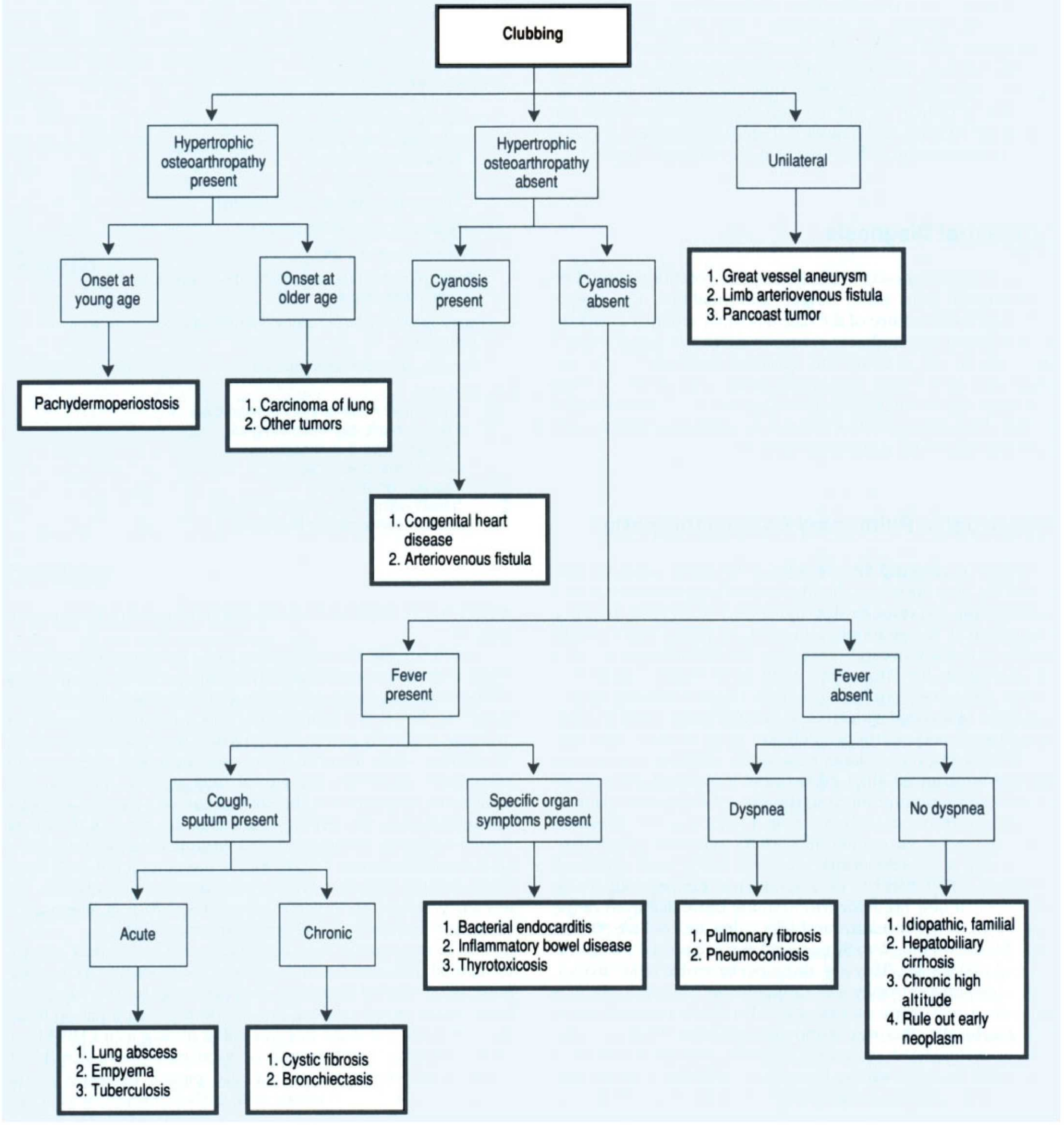

Figure 116-2 Algorithm for the diagnostic evaluation of clubbing.

planes. Also distinctive is an increased mobility of the nail on its bed such that the affected nail can be easily rocked by applying alternating pressure to the base and tip. (A degree of experience is required to distinguish pathologic from normal "rocking" motions.)

Etiology

The etiology of clubbing is always determined by precisely defining the causative clinical disorder (Fig. 116-2). In general, the degree of clubbing parallels the severity of the pulmonary disease; it may abate if the underlying disorder regresses. It is usually symmetric in distribution, but in certain congenital heart diseases the process is asymmetric. For example, only toe clubbing is noted in patent ductus arteriosus with a shunt reversal. Interestingly, unilateral clubbing occurs with aneurysms of the great thoracic vessels (e.g., subclavian artery, aorta), arteriovenous fistulae in the affected limb, ipsilateral Pancoast tumors, hemiplegia, and trauma. Tuberculosis, unless associated with empyema or bronchiectasis, is not a common cause.

Differential Diagnosis

Because of its diagnostic implications, this condition should be differentiated from other finger abnormalities: (1) simple "breaking" or curvature of the nail, a normal variant; (2) Heberden's osteoarthritic nodes; (3) chronic infectious digital arthritis with periarticular swelling and a normal nail bed; (4) chronic paronychia and felons; (5) epidermoid cysts of the osseous phalanges; (6) acromegalic bony enlargement; (7) posthemiplegic digital atrophy; and (8) acrosteolysis, a pseudoclubbing seen in people who work with vinyl chloride.

Hypertrophic Pulmonary Osteoarthropathy

Secondary or acquired hypertrophic pulmonary osteoarthropathy (HPO), also known as the Marie-Bamberger syndrome, is a distinct entity and must be distinguished from simple clubbing, with which it is commonly confused. Acquired HPO usually occurs in certain visceral disorders, but bronchogenic carcinoma accounts for 90 percent of all cases (Table 116-1). Pulmonary metastases rarely cause HPO. Digital clubbing may or may not be associated with HPO, although it is generally present. Digital perfusion patterns are different in these two conditions; in acquired clubbing, blood flow to the digits is augmented, whereas in acquired HPO, blood flow is shunted through arteriovenous communications to the sites of osteoarthropathy.

The syndrome of HPO occurs in 4 to 12 percent of patients with lung carcinoma, commonly with the epidermoid cell type, less so with adenocarcinoma, and only rarely with small cell carcinoma. HPO consists of a symmetric neoperiostitis with subperiosteal new bone formation in the distal diaphysis of the long bones of the forearms and legs (ulna and radius, 80 percent; tibia and fibula, 70 to 80 percent) and occasionally metacarpals and metatarsals. There is also a periarticular inflammation with synovial hypertrophy and joint effusion. The periosteum is raised by a new bone matrix and subsequent mineralization. Radiographically, this new subperiosteal bone formation appears as a thin layer separated from the normal cortex by a radiolucent line. Coexisting features of neurovasomotor instability include episodic swelling and blanching, diaphoresis, and paresthesia of the hands and feet. Pain is the chief clinical complaint as a result of these pathologic changes in the affected ankles, wrists, or tibiae. If digital clubbing coexists, it too is quite painful.

Occasionally, the osteoarthropathy involving wrists, knees, ankles, or elbows presents with swelling, tenderness, morning stiffness, and ankylosing deformities that mimic rheumatoid arthritis. In fact, these manifestations may resemble rheumatoid arthritis so closely that the patient may be erroneously treated for arthritis while the underlying osteoarthropathy remains unrecognized. However, the typical radiographic appearance of long bone periostitis is sufficiently diagnostic, if not essential, to distinguish these two entities. Furthermore, synovial fluid from the affected joints in HPO reveals a noninflammatory fluid that is sparse in leukocytes (<2,000/mm^3) and neutrophils (<15 percent), serving further to differentiate these two conditions. The tendency of this noninflammatory fluid to clot is also a fairly distinctive feature of pulmonary osteoarthropathy.

In some patients, exquisite periosteal tenderness elicited by manual pressure over the involved distal bones is a clue to an underlying remote neoplasm. Radionuclide Tc-99m phosphate bone scans reveal a typical linear isotopic accumulation along the periosteum of distal diaphyses that distinguishes HPO from bony metastases (with central or focal radiotracer density) in cases of carcinoma. Scans may also permit diagnosis of the process in patients presenting with early or vague clinical complaints before the characteristic conventional radiologic changes develop.

Table 116-1 Diseases Associated with Digital Clubbing

- Idiopathic, familial
- Acquired diseases
  - Heart disease
    - Cyanotic (congenital)
    - Bacterial endocarditis
  - Lung disease
    - Carcinoma of lung or pleura
    - Bronchiectasis
    - Lung abscess
    - Empyema
    - Tuberculosis (typically with apical bronchiectasis)
    - Pulmonary fibrosis, pneumoconiosis
    - Cystic fibrosis
    - Alveolar proteinosis
  - Gastrointestinal disorders
    - Regional enteritis
    - Ulcerative colitis
    - Cirrhosis (hepatic, biliary)
    - Amebic dysentery
    - Achalasia, peptic ulcer
    - Hepatic amyloidosis
  - Endocrine disorders
    - Thyrotoxicosis (thyroid acropachy)
  - Any chronic suppurative process
  - Arteriovenous fistulae
  - Rarer: repeated pregnancies, purgative abuse, syphilis, hemiplegic limbs
- Hypertrophic osteoarthropathy (Marie-Bamberger syndrome)
  - Idiopathic
  - Familial pachydermoperiostosis
  - Acquired
    - Lung cancer (primary or metastatic to)
    - Pleural, especially mesothelioma
    - Thymus, thyroid carcinoma
    - Leiomyoma of esophagus
    - Thoracic lymphoma
    - Nasopharyngeal carcinoma
    - Chronic myelogenous leukemia

Primary or Familial Disorders

Secondary (acquired) clubbing and HPO are unrelated to two other rarer, but distinguishable, conditions: pachydermoperiostosis and primary or familial clubbing. Neither disorder is currently known to be associated with any underlying systemic disease.

Pachydermoperiostosis is a familial process (autosomal dominant) characterized by painless clubbing and HPO and particularly coarse facial features with deep furrowing of nasolabial and forehead folds. Hyperhidrosis in these sites is typical. The disorder usually appears at puberty and progresses insidiously for about a decade before becoming stationary. Often, a grotesque deformity of the fingers and toes accompanies the clubbing process. Early osseous changes identical with HPO are limited to the distal portions of long tubular bones, metacarpals, and metatarsals; later, all bones except the skull become affected.

Primary or hereditofamilial clubbing, which may occur in the absence of HPO, appears simply in the first or second decade of life, having been transmitted as an autosomal dominant process with low expression. It is not associated with an increased capillary circulation.

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