2 Cardiac Clues from Physical Appearance

SKIN

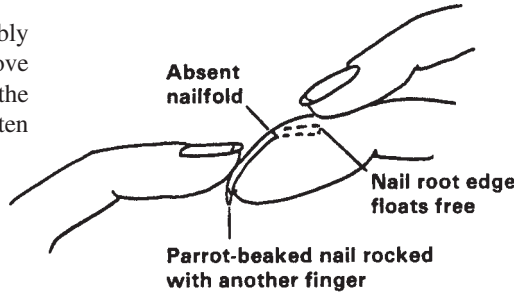
1. What is meant by cyanosis?

   ANS: Cyanosis is the bluish or purplish color imparted to the skin and mucous membranes, usually the result of at least 5 mg per dL of reduced hemoglobin in the surface capillaries, but occasionally due to an abnormal hemoglobin such as sulfmethemoglobin.

2. How can you distinguish central from peripheral cyanosis clinically?

   ANS: Central cyanosis is seen in warm as well as cold areas (compare with the tongue of a normal patient). In African American patients, evidence of central cyanosis may be seen in the conjunctiva. Definite central cyanosis is not usually recognized unless the arterial oxygen saturation is lowered to about 80%. Peripheral cyanosis is seen only in cool areas such as the nail beds, nose, cheeks, earlobes, and the outer surface of lips, where slow flow decreases the amount of hemoglobin in the surface capillaries. If **clubbing** is present or if the hands are warm, the cyanosis is probably central.

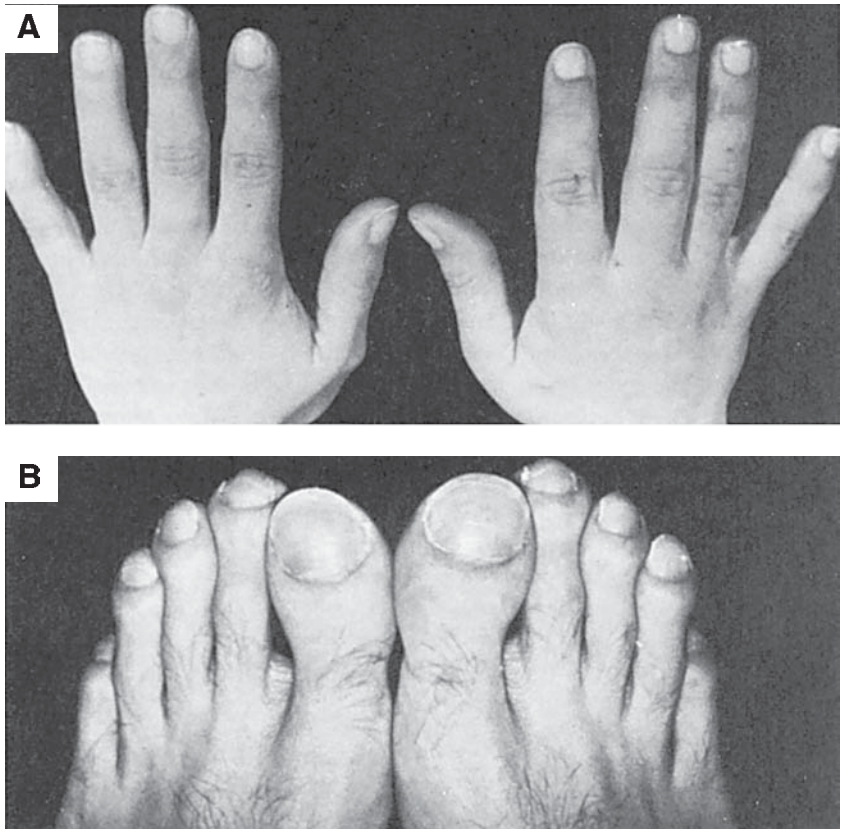
The earliest sign of clubbing is probably the reduction or absence of the groove where the root of the nail slips under the skin. Moist, warm fingertips are often associated signs.



3. How should you test for mild clubbing?

   ANS: a. Look for obliteration of the normal angle between the base of the nail and the proximal skin.

Boldface type indicates that the term is explained in the glossary.



The feet of this 23-year-old man with a reversed shunt through a persistent ductus were cyanotic and clubbed, while his hands were normal.

b. Approach the nail bed from behind and feel the edge of the nail root floating free when the distal portion of the nail is depressed with another finger.

4. What can cause clubbing, besides cyanotic heart disease?

   ANS.: Hypertrophic osteoarthropathy as in cancer of the lung, infective endocarditis, and severe ulcerative colitis.

   Note: Cancer of the lung is the most common cause of painful clubbing.

5. What is differential cyanosis and what is its significance?

   ANS: Differential cyanosis means that the fingers are pink but the toes are cyanotic (and usually clubbed). It signifies the presence of a persistent ductus arteriosus (PDA) with a reversed right-to-left shunt due to pulmonary hypertension (**Eisenmenger syndrome**). The left hand may

also be cyanotic, because the ductus may be located at the branching part of the left subclavian artery.

6. What are the skin signs secondary to the small emboli of **infective endocarditis**?

   ANS: a. Clubbing.
   b. Splinter hemorrhages in the nails. (Most splinter hemorrhages are not embolic and are due to repeated jarring. Since they are in the nail substance, they move with the nail as it grows and they extend to the distal nail edge. Embolic splinters are subungual and usually do not extend to the distal nail edge.) Fresh red hemorrhages, especially at the base of nail, are more important than brown linear streaks near the tips of the fingers. White-centered petechiae are most easily seen on the conjunctiva of the everted lower lid.
   c. Osler's nodes (painful, tender, reddish-brown raised areas 3–15 mm in diameter, occasionally with a whitish center, on the palms or soles).
   d. Janeway lesions (painless, circular or oval, pink to tan macules about 5 mm in diameter on the palms and soles that do not blanch with pressure).

7. What cardiac condition is suggested by brownish, muddy pigmentation of the skin and signs of hepatic failure, such as loss of axillary and pubic hair?

   ANS: Hemochromatosis with a cardiomyopathy due to intracellular iron deposits in the heart muscle, with secondary interstitial fibrosis.

8. When can amyloid disease with cardiac involvement cause skin lesions? What kind of lesions?

   ANS: Only if it is primary or if it is secondary to multiple myeloma, i.e., not if it is secondary to infection. Yellowish brown papules, nodules, or plaques may develop, often pruritic, with signs of bleeding due to scratching.

9. What is livedo reticularis? What is its significance?

   ANS: The term indicates a marbling reticulation or fishnet type of mottling of the lower trunk, buttocks, and extremities, precipitated or exaggerated by cold or by emotional upsets. It occurs in about 20% of patients with lupus erythematosus, periarteritis nodosa, or cryoglobulinemia. If it has occurred recently in a man over age 50, it suggests cholesterol embolization from an abdominal aortic aneurysm [1].

10. What are the cutaneous manifestations of intestinal **carcinoid** disease? What are the cardiac manifestations?

    ANS: The face and neck may show various blends of red and purple, often patchy and mottled. If the disease has been present for a long time, telangiectasia may be present, as may pulmonary or tricuspid stenosis and regurgitation caused by fibrosis of valve structures.

11. What diagnosis can be made by noting facial flushing following a syncopal attack?

    ANS: A cardiac arrest or a **Stokes-Adams attack**. The flush is probably a manifestation of the total-body reactive hyperemia that follows a temporary cessation of circulation.

12. What conditions that can affect the heart, besides thyrotoxicosis, can cause warmer-than-average skin?

    ANS: Severe anemia and beer drinkers' acute **beriberi**.

13. What kind of conditions are suggested by cold hands and feet?

    ANS: a. If hands and feet are moist, they suggest anxiety and may explain chest pains, palpitations, and fatigue as seen in **neurocirculatory asthenia** (DaCosta's syndrome).

    b. If only the feet are cold and the patient has a history of **intermittent claudication**, peripheral arterial obstruction with poor collateral circulation is suggested.

    c. If cold extremities are relatively recent in onset (a few weeks to a few years), a low output state is suggested.

    Note: The cold hands of the low output state can become warm when palmar erythema (liver palms) develops, secondary to cardiac cirrhosis.

14. What are the skin signs of pheochromocytoma of adrenals?

    ANS: Neurofibromatosis, cafe-au-lait spots, and axillary freckling (Crowe's sign).

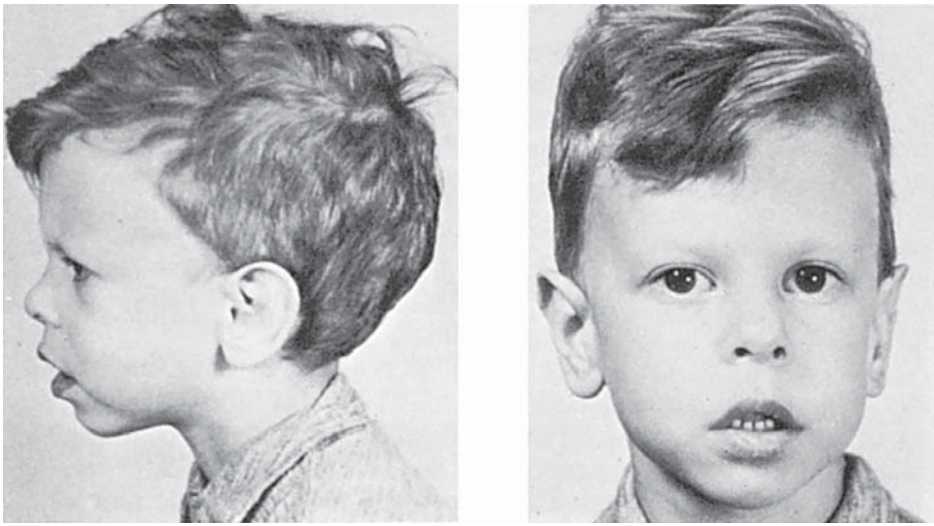
CONGENITAL AND ACQUIRED FACIES

1. What are the facies of supravalvular aortic stenosis, also known as Williams syndrome?

    ANS.: A flattened occiput, broad high forehead, puffy cheeks, low ears, ocular **hypertelorism** with strabimus, underdeveloped nasal bones, an upturned nose with a long filtrum (the vertical groove between nose and mouth), a wide pouting mouth, dental abnormalities, and hypoplastic mandible. Patients commonly have atrial and/or ventricular septal defects.

2. What are the facies of Down syndrome (trisomy 21)? What are the cardiac abnormalities?

    ANS: A flattened occiput, disproportionately small head, epicanthal folds that give the impression of slanted eyes, and a mouth held open by a large protruding tongue. The most common cardiac abnormalities are various combinations of **atrial septal defects**, **ventricular septal defects**,



This boy with severe supravalvular aortic stenosis does not have all its facial characteristics; e.g., there is no hypertelorism or strabismus.

and atrioventricular (AV) valve regurgitation due to various degrees of **endocardial cushion defects**.

3. What is de Musset's sign?

   ANS: Head-nodding movements secondary to the ballistic force of severe aortic regurgitation (AR).

   Note: The sign was named after a patient, Alfred de Musset, a French poet whose nodding movements were described by his brother in a biography.

4. What are the facies of myxedema? What cardiac abnormalities are expected?

   ANS: Puffy lids and loss of the outer third of the eyebrows: scanty, dry, hair; coarse, dry skin; expressionless face, and an enlarged tongue.

   Note: These patients have cardiomyopathies due to increased interstitial fluid and mucoid infiltration. They also have pericardial effusions.

5. What is meant by an earlobe crease, and what is its significance?

   ANS: This is an oblique crease in the earlobe. Ninety percent of patients over age 50 with significant triple vessel coronary disease have a deep earlobe crease. A unilateral ear crease was found in one study to be associated with an intermediate degree of coronary obstruction [2]. (See p. 22.)



This 47-year-old man had this deep ear crease bilaterally. Although he had no significant coronary disease, his cholesterol-to-HDL ratio was 8 to 1, and he had sinus node dysfunction. He was about 50 lb overweight.

6. What is the malar flush?

   ANS: Cyanotic cheeks with a slight telangiectasia. It was once thought to be specific for mitral stenosis, pulmonary hypertension, and high venous pressure. However, it can be seen in any patient with low cardiac output and high venous pressure. It can also be seen in those who lead an outdoor life, as well as in patients with myxedema. It is sometimes seen in patients with severe pulmonary stenosis with atrial septal defects and cushingoid moon facies, as well as in the carcinoid syndrome.

   In systemic lupus, the cheeks will also be reddened, but with a butterfly erythema that covers the cheeks and bridge of the nose. In these patients lesions are also found in exposed areas such as the scalp, external auditory canal, neck, and upper chest. The heart may be involved in lupus with pericarditis, myocarditis, Libman-Sacks verrucous endocarditis of the mitral or aortic valve, and occasional myocardial infarction due to coronary arteritis.

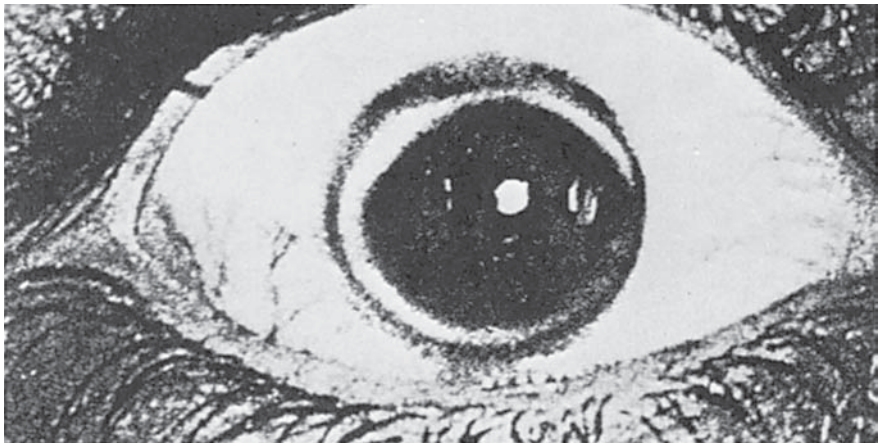
EYES

1. What is the cardiac cause of jaundice?

   ANS: Jaundice may be a sign of severe heart failure, with high venous pressure causing liver damage.

2. Which kind of corneal arcus (circumferential light gray or yellowish ring around the rim of the iris) is associated with hypercholesterolemia or coronary disease?

   ANS: A thick band that begins inferiorly and is inside the limbus, allowing a thin rim of iris pigment to be seen between the arcus and the sclera. The



This type of arcus is a thick band of yellowish material surrounded by peripheral pigment and suggests a high serum cholesterol. It is not an arcus senilis, which has little known significance. (Courtesy Ayerst Laboratories.)

usual "arcus senilis" is not necessarily associated with hyperlipidemia or coronary disease. It begins superiorly and extends to the rim or limbus of the iris.

3. How does **infective endocarditis** affect the eyes?

   ANS: a. Conjunctival hemorrhages and petechiae (due to tendency to bleed plus minute emboli). Evert lids to see these lesions.
   b. Oval or canoe-shaped hemorrhages near optic disk with white spot in center (Roth spots).

4. With which cardiac lesions are cataracts associated?

   ANS: They may be part of the rubella syndrome in which a PDA and pulmonary artery (not valve) stenosis are the most common cardiac lesions. Other features may include deafness and mental deficiency due to microcephaly. The rate of growth may be slow.

5. Which cardiac lesion should you suspect in the presence of an Argyll Robertson pupil (reacts to accommodation but not to light)?

   ANS: Luetic aortic aneurysm or luetic AR with coronary ostial stenosis.

6. Which cardiac condition besides thyrotoxic heart disease may be associated with exophthalmos?

   ANS: Advanced congestive heart failure with high venous pressure and weight loss. The stare is probably due to lid retraction caused by the strong sympathetic tone accompanying the low cardiac output, and exaggerated by the slight proptosis.

7. What cardiac lesions should you suspect if you see a tremulous iris (iridodonesis) when gaze shifts rapidly from side to side?

   ANS: This sign suggests the Marfan syndrome, in which the iris is not properly supported by the lens because of dislocation or weakness of the suspensory ligament. The cardiac lesions associated with it are aneurysms of the aorta or pulmonary artery and myxomatous degeneration of the aorta or mitral valve, with consequent regurgitation.

8. What is the significance of xanthelasma (flat **xanthomas** or yellowish cholesterol-filled plaque on or around the eyelid)?

   ANS: It is usually associated with hypercholesterolemia.

9. What are the retinal signs of various degrees of **arteriosclerosis** [3]?

   ANS: Grade 1: The light reflex is increased in width.
   Grade 2: Crossing abnormalities (arteriovenous nicking and right-angled crossing of the arteries over the veins).
   Grade 3: Copper-wire arteries (red color of artery is slightly brownish due to thick walls).
   Grade 4: Silver-wire arteries (no red color is seen, only a whitish light reflex).

   Note: Hollenhorst plaques are flakes of cholesterol emboli seen as glinting spots, often seeming larger than the vessels in which they reside.

10. What are the retinal signs of different degrees of hypertension?

    ANS: Grade 1: Generalized attenuation (arteriovenous ratio of less than 2 to 3).
    Grade 2: Focal constriction or spasm.
    Grade 3: Hemorrhages and exudates. (Exudates may either resemble cotton wool or be hard and shiny.)
    Grade 4: Papilledema.

    Note: Pure attenuation of the arterioles is best seen in toxemia of pregnancy or in young persons with rapid onset of hypertension. Minimal narrowing is most easily seen beyond the first or second bifurcation where the arteries actually become arterioles. Corkscrew tortuosity with frequent "U turns" may be seen with coarctation and sometimes with retinal hemorrhages.

11. What three cardiac conditions are associated with a blue sclera?

    ANS: a. Osteogenesis imperfecta is associated with AR.
    b. The Marfan syndrome is associated with great-vessel aneurysms and mitral or aortic valve regurgitation.

c. Ehlers-Danlos syndrome, with its hyperelastic, fragile skin, hyperextensible joints, and kyphoscoliosis, is associated with atrial septal defect (ASD), tetralogy of Fallot, or regurgitant valves.

12. What are angioid streaks?

ANS: Brown linear streaks perpendicular to blood vessels. Seen in pseudoxanthoma elasticum and endocardial fibrosis and regurgitation in the heart.

EDEMA

1. How can you demonstrate edema even when it is slight?

ANS: Press on the skin over a bony area for 10 s with at least three fingers spread slightly apart, and feel for valleys between hills after release.

2. What is the significance of slow and fast edema?

ANS: With slow edema, the pitting remains for more than 1 min, and it is most likely due to congestion. If, however, the pitting disappears in less than 40 s (fast edema), the cause is almost certainly a low albumin level [4]. (The venous pressure presumably controls the rate of tissue fluid flow from the legs.)

3. What are the usual causes of noncardiac bilateral leg edema, besides low albumin?

ANS: a. Premenstrual edema (hormonal).

b. Tight undergarments.

c. Obesity (obstructed lymphatics).

4. How can you rule out a cardiac cause for the edema?

ANS: Check the venous pressure. Normal venous pressure is incompatible with a cardiac cause of the edema unless diuretics have been given. Face and hand edema tend to rule out a cardiac cause except in infants.

5. Where should you check for edema in a bedridden patient?

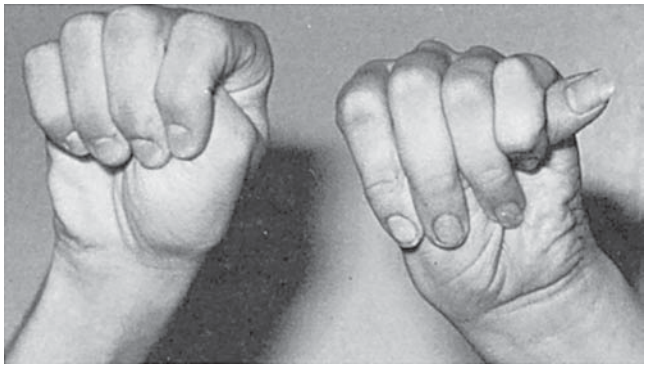
ANS: Only presacral edema may be present if the patient has been in bed for some time.

6. How much body fluid may collect in tissue before pitting occurs?

ANS: At least 10 lb (4.5 kg).

7. How can you tell that ascites is present?

ANS.: If flanks are resonant, swelling is probably gas, common in severe congestive heart failure (CHF). If ascites is from CHF, the liver is enlarged and often ballotable.



At left is a normal subject, who is unable to protrude his thumb beyond his clenched fingers, as can the patient with the Marfan syndrome at right, who can do this because of a long thumb and lax joints.



The normal patient at left cannot overlap his thumb and little finger around his wrist because, unlike the patient with the Marfan syndrome at right, his fingers are not long relative to his wrist.

EXTREMITIES

1. What hand, wrist, and extremity findings suggest the Marfan syndrome, with its possible cardiac abnormalities of aortic regurgitation and prolapsed mitral valve?

   ANS:
   a. Fingers: slender and long ("spider" fingers, or arachnodactyly).
   b. Thumb sign: when a fist is made over a clenched thumb, the thumb should not extend beyond the ulnar side of the hand. (False positives of this sign occur in 1% of white children and 3% of African American children.)
   c. Wrist sign: when the wrist is encircled by the thumb and the little finger (with light pressure), the little finger will overlap at least 1 cm in 80% of patients with the Marfan syndrome.
   d. The span of the outstretched arms exceeds the height by at least 5 cm.

2. What is the most common cardiac lesion associated with rheumatoid arthritis?

   ANS: Pericarditis and even occasional constrictive pericarditis.

3. What skeletal abnormalities suggest an atrial septic defect (ASD)?

   ANS: a. Any skeletal deformity of the Marfan syndrome suggests not only pulmonary artery or aortic dilatation but also an ASD.

   b. A prominent left precordium suggests not only that the right ventricle (RV) was dilated during childhood but also that it was working against a high pressure. This deformity suggests that the ejection murmur is due to an ASD with hyperkinetic pulmonary hypertension.

   c. A thumb deformity such as a fingerlike thumb (three phalanges) or an extra-short thumb, combined with an ASD, is called the Holt-Oram syndrome. An ulnar-radial deformity that prevents good forearm supination or pronation may be present with this syndrome.

4. What cardiovascular abnormality is associated with Turner's syndrome?

   ANS: Coarctation.

5. What are the hand signs of Down's syndrome, besides the simian crease?

   ANS.: The fourth and fifth fingers are abnormally separated and the fifth finger is short and curved inward.

CHEST AND RESPIRATION

1. Which cardiac abnormalities are suggested by a **pectus excavatum**?

   ANS: a. This may occur in the Marfan syndrome, with aortic or pulmonary artery aneurysms, or myxomatous degeneration of the mitral or aortic valve with regurgitation. An ASD should also be suspected. (Pectus carinatum, or pigeon breast, may also occur with the Marfan syndrome.)

   b. It may be part of the straight-back syndrome (see Chapter 13).

2. What should **Cheyne-Stokes respiration** suggest in a cardiac patient?

   ANS.: A very low output at rest. The anoxic respiratory center becomes insensitive to normal tensions of CO_2. The apnea causes hypercapnea, which stimulates the respiratory center, resulting in hyperpnea, which washes out the CO_2 and apnea recurs.

3. What is the significance of a short breath-holding time?

   ANS: It signifies either

   a. Chronic hyperventilation, or

   b. Poor psychophysical control of the breathing apparatus. The normal subject (with legs dangling) can hold his or her breath for 30 s

with a little encouragement. Inability to hold it for at least 20 s is abnormal and can explain dyspnea on exertion. Heart failure or chronic obstructive pulmonary disease (COPD) does not shorten breath-holding time unless there is dyspnea at rest.

4. What is a shield chest, and what does it suggest?

   ANS: It is a broad chest with a greater angle than usual between the manubrium and the body of the sternum, as well as widely separated nipples. In a female with neck webbing, wide carrying angle, and short stature (under 5 ft in height) it suggests **Turner's syndrome** and concomitant coarctation. In a male it is called Noonan's or Ullrich's syndrome and is commonly associated with pulmonary stenosis.

5. What is Ewart's sign of a large pericardial effusion?

   ANS.: Compression of the lung results in a patch of marked dullness between the angle of the left scapula and the spine, associated with tubular (bronchial) breathing and egophany in the same area.

6. How can crackles (formerly called rales) help diagnose lung abnormalities?

   ANS.: Paninspiratory: pneumonia or CHF. Early: severe COPD, asthma or chronic bronchitis. Late: interstitial edema, pulmonary fibrosis, or resolving CHF. Early to mid: bronchiectasis.

   Note: Most crackles are not due to CHF but due to sputum in the air passages which often disappear with several coughs. In early CHF crackles may be absent.

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