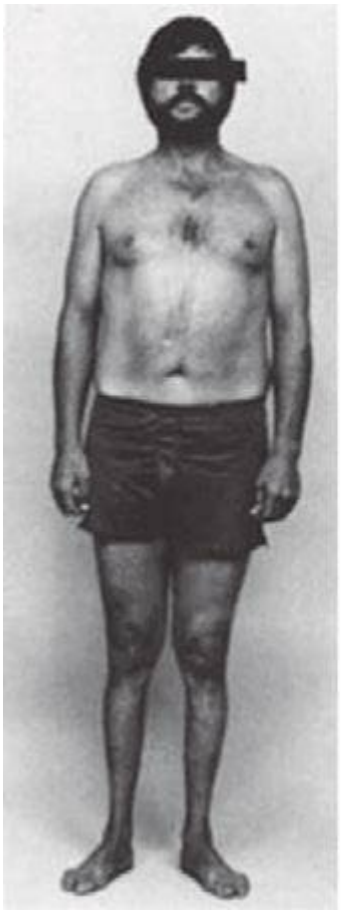


FIGURE 436e-19 Progressive myopathy in a patient with type IIIa glycogen storage disease. The patient has a debrancher deficiency in both liver and muscle (subtype IIIa). As a child, he had hepatomegaly, hypoglycemia, and growth retardation. After puberty, he no longer had hepatomegaly, and his final height is normal. Note the muscle wasting in the lower legs and both hands at age 44 years of age (*left panel*); this condition progressed to pronounced muscle atrophy at age 53 years (*two right panels*). *(Source: CR Scriver et al [eds]: The Metabolic and Molecular Bases of Inherited Disease online, 8th ed. New York, McGraw-Hill, www.ommbid.com.)* **See Chap. 433e.**

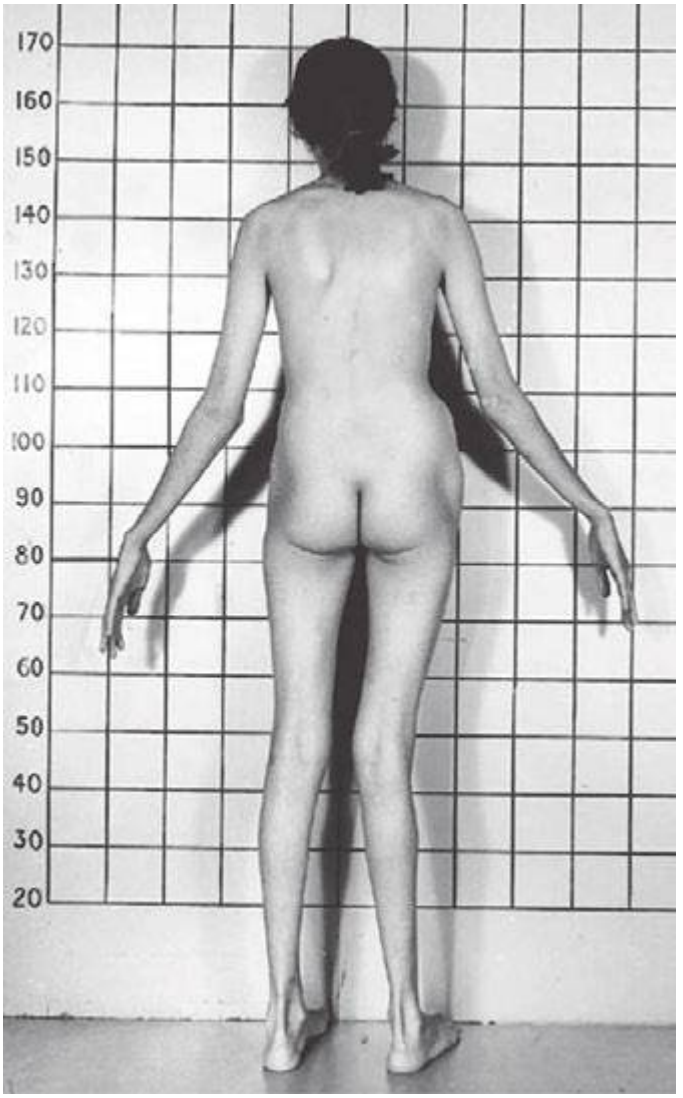
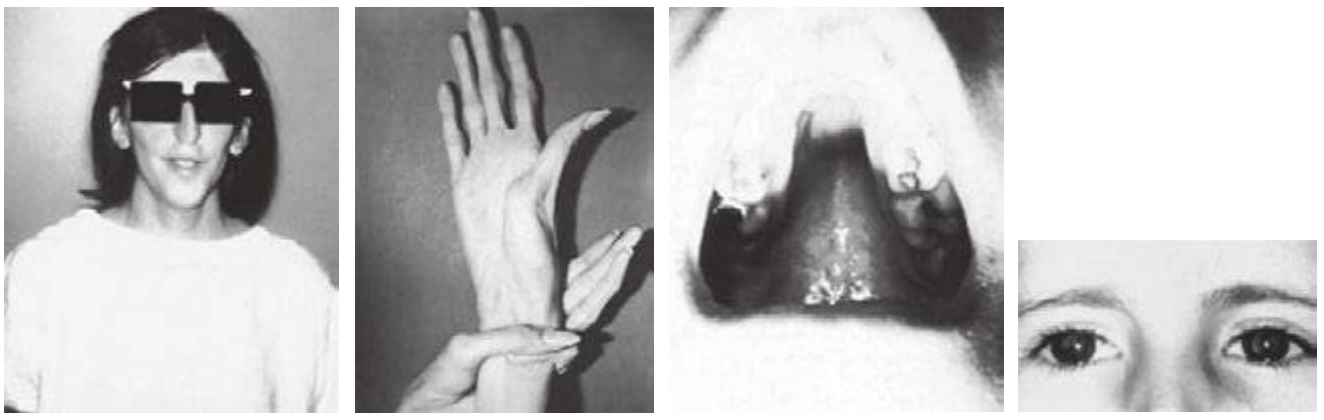


FIGURE 436e-20 Skeletal features of Marfan's syndrome in a 16-year-old girl. Note the long limbs (associated with disproportionately tall stature), long fingers, scoliosis, and genu valgum. *(Source: CR Scriver et al [eds]: The Metabolic and Molecular Bases of Inherited Disease online, 8th ed. New York, McGraw-Hill, www.ommbid.com.)* **See Chap. 427.**



A ***B*** ***C*** ***D***

FIGURE 436e-21 Marfan's syndrome. ***A.*** Long, narrow face. ***B.*** Arachnodactyly and positive wrist sign. ***C.*** High-arched palate. ***D.*** Ectopia lentis associated with aortic aneurysm and severe aortic regurgitation in a teenage girl. *(Source: V Fuster et al [eds]: Hurst's The Heart, 11th ed. New York, McGraw-Hill, 2004, www.accessmedicine.com.)* **See Chap. 427.**