

# Klippel-Trénaunay syndrome

## Klippel-Trénaunay syndrome

This is a combined anomaly of a cutaneous naevus, persistent vestigial veins with varicose veins and soft-tissue and bone hypertrophy. The condition is a mesodermal abnormality that is not familial ( Figure 62.36 ). Segments of the deep veins are hypoplastic or aplastic and there may be an associated obstruction of the lymphatics. The condition must be distinguished from the Parkes-Weber syndrome, in which there are multiple arteriovenous fistulae causing venous hypertension, ulceration and high-output cardiac failure. Virtually all patients with Klippel-Trénaunay syndrome should be treated conservatively with compression hosiery; however, some will benefit from laser ablation of the naevus, stapling of the bones to avoid leg length discrepancy and occasional removal of large superficial varicose veins, provided the

George Budd, 1808–1882, Professor of Medicine, King's College Hospital, London, UK, described this syndrome in 1845. Hans Chiari, 1851–1916, Professor of Pathological Anatomy, Strasbourg, Germany (Strasbourg was returned to France after the end of the First World War, in 1918), gave his account of this condition in 1898. Frederick Parkes-Weber, 1863–1962, physician, The German Hospital, Dalston, London, UK. Sir James Paget, 1814–1899, English surgeon and pathologist, best known for his description of Paget's disease of the bone. Leopold von Schrotter, 1837–1908, Austrian physician and laryngologist, Chair of Laryngology, University of Vienna, Vienna, Austria. deep veins are patent. LMWH should be given to all patients having surgery as this syndrome is associated with an increased risk of VTE. -

Figure 62.36 Two patients with Klippel-Trénaunay syndrome. (a) This patient has a longer leg and a capillary naevus; (b) this patient has a large lateral anomalous axial vein.

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