Patients with Turner syndrome (TS) have increased fracture risk and decreased bone density, patients with severe hemophilia have low bone density. The etiology of these changes hasn't been completely elucidated in any of the two diseases. Our aimes were to assess bone density and geometry at the radius using a new method peripheral quantitative CT densitometry (pQCT) and to describe associations between densitometry parameters and estrogen treatment in TS and laboratory as well as clinical markers of disease severity in haemophilia. Sixty-seven girls with TS (median age 14.3 years, range 6.0-19.4) and 42 boys with haemophilia (median age 12.7 years, range 6.6-19.2) have been measured using peripheral quantitative CT at the radius. The results have been compared to published reference data. Girls with TS had decreased cortical bone density and thinner cortex. These parameters were positively correlated to the length of estrogen treatment. Trabecular bone density was normal in prepubertal girls but it was decreased after puberty. There was no association between trabecular bone density and the estrogen treatment. Boys with haemophilia had decreased trabecular density and low muscle area at the forearm. Densitometry parameters were influenced neither by the clotting factor VIII/IX level nor by the frequency of intra-articular and intramuscular bleeding. While the decrease in bone density in girls with TS is probably caused by estrogen deficiency, the primary cause of bone deficit in boys with haemophilia is sarcopenia.