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7 Association of novel
mutations in CYP46A1
with childhood X-linked
steroid sulfatase
deficiency. Steroid
sulfatase (STS)
deficiency is an X-linked
recessive disorder of
childhood caused by
mutations in the STS

gene, which may be due to point mutations in the gene coding sequences, deletions, or intragenic recombination. Twelve patients with STS deficiency were found to harbor two novel mutations on the same allele in the CYP46A1 gene and were identified by PCR-based sequence analysis. Seven patients had

mutations identified on the CYP46A1 gene (six point mutations and a deletion of 23 bp). Two children carried a mutation in the CYP46A1 gene (one point mutation and a recombinant fragment), whereas the remaining three cases presented with the same mutation on the CYP46A1 gene (two point mutations and a deletion of 23

bp). These data raise the possibility that the CYP46A1 gene may be involved in the development of some cases of STS deficiency. Trans-tibial single-stage repair of congenital dislocation of the tarsal navicular: a technique of choice. A single-stage trans-tibial repair of a dislocated tarsal navicular was performed in a group of

12 feet in 11 patients. The mean age at operation was 2.5 years. The talar dome was reattached to the tarsal navicular and the talonavicular joint was maintained with repair of the capsular and syndesmotic ligaments. All 12 feet had full range of motion in the subtalar joint and no pain. One patient had a wound infection. The

authors recommend that this procedure be considered the method of choice for stabilization of the talonavicular joint in patients with congenital dislocation of the tarsal navicular.

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