**Abstract**

PURPOSE: To describe ocular manifestations in children with congenital insensitivity to pain with and without anhidrosis (CIPA and CIP).

DESIGN: Retrospective observational case series of 39 children.

METHODS: Collected data included demographic information, medical history, ocular surgeries, genetic analysis, and ocular examination results including visual acuity, cycloplegic refraction, ocular surface findings, corneal sensitivity, tear film production, and fundoscopy results. Corneal tomography and slit-lamp photography were performed for some patients.

RESULTS: CIPA was diagnosed in 32 children and CIP in 7: 11 CIPA patients had mutations in *NTRK1* (1926 T ins), 3 CIP patients had mutations in *PRDM12*, and another 3 in *SCN9A*. Mean follow-up time was 49.0 months for CIPA and 85.2 months for CIP. Corneal opacities were seen in 31% of CIPA eyes and 85% with CIP. Most CIPA patients had no superficial punctate keratopathies (SPKs), whereas SPKs were marked in 66% of CIP eyes. Schirmer was normal in 83% of CIPA eyes but reduced in 75% of CIP eyes. Corneal reflex was present in 52% of CIPA eyes and 33% of CIP eyes. Visual acuity was ≥20/25 in most CIPA patients, but 20/30 in 5 and ≤20/40 in 9 CIP eyes.

CONCLUSION: Children with congenital insensitivity to pain are prone to ocular complications and primary corneal scarring. CIP patients tend to have more severe ocular surface disease than those with CIPA, likely due to more pronounced loss of corneal sensation. Affected children should have routine follow-up to identify ocular surface disease and prevent vision loss.