

**University of Illinois at Chicago
Department of Ophthalmology and Visual Sciences
New Research Publications by Month**

April 2006

Cogan's syndrome: a cause of progressive hearing deafness.

American Journal of Otolaryngology

In 1934 Morgan and Baumgartner first described a nonsyphilitic interstitial keratitis (IK) associated with vestibuloauditory dysfunction (Morgan RF, Baumgartner CF, Menier's disease complicated by recurrent interstitialkeratitis. Excellent result following cervical ganglionectomy. *West J Surg* 1934;42:628). Cogan was the first to describe this syndrome as a clinical entity with the report of 5 additional cases in 1945 (Cogan DA: Syndrome of nonsyphilitic interstitial keratitis and vestibuloauditory symptoms. *Arch Ophthalmol* 1945;33:144-9). Since that time, more than 100 cases of Cogan's syndrome (CS) have been in the literature. Cogan's syndrome is a rare clinical disease, which primarily affects young adults; however, published reports range from 2.5 to 60 years for age of onset (Kundell Sp, HD Ochs: Cogan's syndrome in childhood. *J Pediatr* 1980;97:96-8). This disease primarily affects whites and is believed not to be hereditary. Typical CS is characterized by IK and vestibuloauditory dysfunction. The IK usually occurs with sudden onset and is characterized by photophobia, lacrimation, and eye pain. The vestibuloauditory dysfunction is usually bilateral, presenting with tinnitus, sensorineural hearing loss, and acute episodes of vertigo. Atypical CS presents with significant inflammatory eye disease (ie, scleritis, episcleritis, retinal artery occlusion, choroiditis, retinal hemorrhages, papilloedema, exophthalmos, or tenonitis) with or without IK (*Laryngoscope* 1960;70:447-9). In this report, we describe a typical case of CS, in which the hearing loss was unresponsive to corticosteroid therapy.

Cundiff, Jason. Kansal, Sukesh. Kumar, Arvind. [Goldstein](#), Debra A. [Tessler](#), Howard H. Cogan's syndrome: a cause of progressive hearing deafness. *Am J Otolaryngol*. 2006 Jan-Feb;27(1):68-70. PMID: 16360829

Rho-associated protein kinase inhibitor, Y-27632, induces alterations in adhesion, contraction and motility in cultured human trabecular meshwork cells.

Experimental Eye Research

We investigated the roles of Rho-associated protein kinase (ROCK) in regulating activities such as adhesion, contraction and migration in cultured human trabecular meshwork (TM) cells. Human TM cells in culture were treated with Y-27632, a specific ROCK inhibitor. Trypan blue exclusion test and TUNEL staining showed little or no direct toxicity of Y-27632 on TM cells. By MTT assay, Y-27632 did not significantly affect the proliferation of TM cells. The cell adhesion assay showed that Y-27632 promoted the cell adhesiveness to both fibronectin and collagen type I in a dose-dependent manner. Collagen gel contraction activity of TM cells was significantly inhibited by the treatment of Y-27632 in a dose-dependent manner. The addition of Y-27632 accelerated motility of TM cells in wound healing assay. Phosphorylated LIM kinase 2 and cofilin, related to actin bundling and integrin clustering, were dephosphorylated (activated) by Y-27632. In conclusion, Y-27632 elicits profound effects on TM cell activities including adhesion, gel contraction, and cell motility. These

Y-27632-induced changes of TM cells may be relevance to the physiology of the aqueous outflow system.

Koga, Tomoyo. Koga, Takahisa. Awai, Maiko. Tsutsui, Jun-ichiro. [Yue](#), Beatrice Y J T. Tanihara, Hidenobu. Rho-associated protein kinase inhibitor, Y-27632, induces alterations in adhesion, contraction and motility in cultured human trabecular meshwork cells. *Exp Eye Res.* 2006 Mar;82(3):362-70. Epub 2005 Aug 25.
PMID: 16125171

Immunolocalization of CYP1B1 in normal, human, fetal and adult eyes.

Experimental Eye Research

CYP1B1 is a cytochrome P450 enzyme implicated in autosomal recessive primary congenital glaucoma (PCG). The mechanism and function of CYP1B1 in the development of the PCG phenotype is unknown. Previously, investigators have reported detection of Cyp1b1 mRNA in the ciliary body and epithelium and neuroepithelium in the developing mouse eye, employing in situ hybridization techniques. Similarly, additional investigators have detected CYP1B1 mRNA in the iris, ciliary body, non-pigmented ciliary epithelial line, cornea, retinal-pigment epithelium, and retina in the human adult eye, using Northern blotting. This study was designed to immunolocalize CYP1B1 protein in the various ocular structures of normal, human fetal and adult eyes. Normal fetal and adult eyes were immunolabeled with a polyclonal antibody against human CYP1B1 using indirect immunofluorescence, and then compared with appropriate controls. The intensity of immunolabeling of the various ocular structures was assessed by qualitative and semi-quantitative techniques. In the anterior segment anti-CYP1B1 immunoreactivity (IR) was detected early in fetal development in the primitive ciliary epithelium. As well, the most intense CYP1B1 IR was in the non-pigmented ciliary epithelium. In addition, CYP1B1 IR was also present in the corneal epithelium and keratocytes, both layers of the iris pigmented epithelium, and retina. However, CYP1B1 IR was absent in the trabecular meshwork in all of the samples. In general, CYP1B1 immunolabeling in the human fetal eyes was more intense when compared to adult eyes. CYP1B1 IR was primarily immunolocalized to the non-pigmented ciliary epithelium and early in fetal development. In addition, CYP1B1 IR was not detected in the trabecular meshwork. These findings suggest that the abnormalities in the development of the trabecular meshwork in PCG may result from diminished or absent metabolism of important endogenous substrates in the ciliary epithelium due to non-functional CYP1B1 enzyme.

Doshi, Manali. Marcus, Craig. Bejjani, Bassem A. [Edward](#), Deepak P. Immunolocalization of CYP1B1 in normal, human, fetal and adult eyes. *Exp Eye Res.* 2006 Jan;82(1):24-32. Epub 2005 Jun 24.
PMID: 15979611

Clinical profile and early surgical complications in the Cornea Donor Study.

Cornea

PURPOSE: The Cornea Donor Study was designed to investigate the safety and efficacy of older donor corneal tissue compared with younger donor tissue in recipient eyes at moderate risk to the graft from progressive endothelial failure. Baseline patient data, including indications for transplant, intraoperative complication rates, and early postoperative complication rates are described herein. **METHODS:** This study was a multicenter prospective, double-masked, controlled clinical trial.

RESULTS: Fuchs dystrophy was the most common indication for corneal transplantation (61%). Intraoperative complications occurred in 33 (3%) patients. A persistent epithelial defect was the most commonly reported postoperative complication, occurring in 92 patients (8%).

CONCLUSION: Intraoperative and postoperative complication rates were low. There was no apparent association between donor or recipient age and either intraoperative or early postoperative complication rates.

Mannis, Mark J. Holland, Edward J. Beck, Roy W. Belin, Michael W. Goldberg, Marc A. Gal, Robin L. Kalajian, Andrea D. Kenyon, Kenneth R. Kollman, Craig. Ruedy, Katrina J. Smith, Patricia. [Sugar](#), Joel. Stark, Walter J. Cornea Donor Study Group. Clinical profile and early surgical complications in the Cornea Donor Study. *Cornea*. 2006 Feb;25(2):164-70.

PMID: 16371775

Attainment of educational levels in patients with Leber's congenital amaurosis.

Ophthalmology

PURPOSE: To assess the educational level attained by patients legally blind with Leber's congenital amaurosis (LCA). DESIGN: Cross-sectional assessment. INTERVENTION: None. MAIN OUTCOME MEASURE: Highest educational level attained by 55 patients with LCA. RESULTS: A total of 55 patients with LCA were included in the study. Of the 55, 54 finished high school. In addition, 36 patients (65%) completed a college education and received a bachelor's degree, and 5 additional patients (9%) were recently accepted to college, whereas 3 others (5%) were currently attending college classes. Further, 18 patients were either pursuing (n = 3) or had attained (n = 15) an educational level beyond a bachelor's degree. CONCLUSIONS: Compromised visual function does not preclude the successful attainment of an academic education in patients with LCA who are substantially visually impaired from birth. These data have clinically relevant implications for the parents of children with LCA and for the patients themselves in providing a tone of optimism for their potential of attaining competitive academic achievements.

PMID: 16513462 [PubMed - indexed for MEDLINE]

[Apushkin](#), M A. [Fishman](#), G A. Attainment of educational levels in patients with Leber's congenital amaurosis. *Ophthalmology*. 2006 Mar;113(3):481-2.