
Because of modern life-support systems capable of keeping tiny premature infants alive, retinopathy of prematurity has recurred. No classification system currently available adequately describes the observations of the disease being made today. A new classification system, the work of 23 ophthalmologists from 11 countries, is presented in an attempt to meet this need. It emphasizes the location and the extent of the disease in the retina as well as its stages. The term "plus" is employed with the stage to denote progressive vascular incompetence. A computer-compatible diagram for recording the results of the examination employing the new classification system is furnished.


We report the preliminary three-month outcome of a multicenter randomized trial of cryotherapy for treatment of retinopathy of prematurity (ROP). Transcleral cryotherapy to the avascular retina was applied in one randomly selected eye when there was threshold disease (defined as five or more contiguous or eight cumulative 30 degrees sectors [clock hours] of stage 3 ROP in zone 1 or 2 in the presence of "plus" disease). An unfavorable outcome was defined as posterior retinal detachment, retinal fold involving the macula, or retrolental tissue. At this writing, 172 infants had been examined three months after randomization. An unfavorable outcome was significantly less frequent in the eyes undergoing cryotherapy (21.8%) compared with the untreated eyes (43%). While the surgery was stressful, no unexpected complications occurred during or following treatment. These data support the efficacy of cryotherapy in reducing by approximately one half the risk of unfavorable retinal outcome from threshold ROP.


OBJECTIVE: To evaluate outcome at 5 1/2 years after randomization in eyes that underwent cryotherapy and in control eyes of patients in the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity. DESIGN: During infancy, patients with bilateral threshold retinopathy of prematurity (ROP) were assigned to receive cryotherapy for one eye and no cryotherapy for the other eye. Those with threshold ROP in only one eye (asymmetric) were randomly assigned to cryotherapy or no cryotherapy for that eye. Then, 5 1/2 years later, testers who were masked to the treatment status of each eye measured the patients' monocular visual acuity by using the log of the minimum angle of resolution visual acuity chart that was used in the Early Treatment of Diabetic Retinopathy Study. This was the most refined visual acuity testing yet performed on this cohort. Structural outcome was evaluated by participating ophthalmologists' assessment of ROP residual in the posterior pole of the fundus. PATIENTS: Patients were 291 children who had been preterm infants with birth weights of less than 1251 g in whom threshold ROP had developed. Two hundred forty patients had bilateral threshold ROP, and 51 had threshold ROP in only one eye. RESULTS: For the 234 children examined, both visual acuity and fundus structure showed a reduction in unfavorable outcomes in treated vs control eyes: 47.1% vs 61.7%, respectively (P<.005), for visual acuity and 26.9% vs 45.4%, respectively (P<.001), for fundus status. Detailed analysis of visual acuity outcomes for all eyes revealed that while fewer treated eyes (31.5%) than control eyes (48%) were blind (P<.001), there was a slight trend toward fewer eyes with a visual acuity of 20/40 or better in the treated (13%) vs control (17%) groups (P=.19). CONCLUSIONS: The results support the long-term efficacy and safety of cryotherapy in the treatment of severe ROP. However, the data show preliminary evidence of a possible adverse effect of this treatment on visual acuity.


BACKGROUND: The LIGHT-ROP Study was a multicenter, prospective, randomized, controlled, clinical trial designed to determine if a reduction in ambient light exposure to premature infants' eyes would reduce the incidence of retinopathy of prematurity (ROP). Previous clinical reports were contradictory and had one or more methodological flaws. METHODS: The LIGHT-ROP trial was designed in rigorous fashion to include randomization to treatment or control groups, masking of examiners, an adequate sample size of high-risk infants, a universal ROP classification system, a reliable and reproducible ophthalmologic examination protocol, a controlled method of light reduction, and monitoring of light levels in the nursery. The multicenter trial was conducted at three centers in Buffalo, Dallas, and San Antonio, with a separate coordinating center in Houston. The trial also utilized a separate data and safety monitoring committee. CONCLUSION: The design of the LIGHT-ROP study represents the most rigorous and largest clinical trial to date. It has demonstrated that light reduction does not reduce the incidence of confirmed ROP in high risk infants.


OBJECTIVE: To determine the efficacy and safety of supplemental therapeutic oxygen for infants with prethreshold retinopathy of prematurity (ROP) to reduce the probability of progression to threshold ROP and the need for peripheral retinal ablation. METHODS: Premature infants with confirmed prethreshold ROP in at least 1 eye and median pulse oximetry <94% saturation were randomized to a conventional oxygen arm with pulse oximetry targeted at 89% to 94% saturation or a supplemental arm with pulse oximetry targeted at 96% to 99% saturation, for at least 2 weeks, and until both eyes were at study endpoints. Certified examiners masked to treatment assignment conducted weekly eye examinations until each eye reached opthalmic endpoint. An adverse opthalmic endpoint for an infant was
defined as reaching threshold criteria for laser or cryotherapy in at least 1 study eye. A favorable ophthalmic endpoint was regression of the ROP into zone I for at least 2 consecutive weekly examinations or full retinal vascularization. At 3 months after the due date of the infant, ophthalmic findings, pulmonary status, growth, and interim illnesses were again recorded. RESULTS: Six hundred forty-nine infants (325 conventional and 324 supplemental) were enrolled from 30 centers over 5 years. Five hundred ninety-seven (92.0%) infants attained known ophthalmic endpoints, and 600 (92%) completed the ophthalmic 3-month assessment. The rate of progression to threshold in at least 1 eye was 48% in the conventional arm and 41% in the supplemental arm. After adjustment for baseline ROP severity stratum, plus disease, race, and gestational age, the odds ratio (supplemental vs conventional) for progression was 72 (95% confidence interval: 52, 1.01). Final structural status of all study eyes at 3 months of corrected age showed similar rates of severe sequelae in both treatment arms: retinal detachments or folds (4.4% conventional vs 4.1% supplemental), and macular ectopia (3.9% conventional vs 3.9% supplemental). Within the prespecified ROP severity strata, ROP progression rates were lower with supplemental oxygen than with conventional oxygen, but the differences were not statistically significant. A post hoc subgroup analysis of plus disease (dilated and tortuous vessels in at least 2 quadrants of the posterior pole) suggested that infants without plus disease may be more responsive to supplemental therapy (46% progression in the conventional arm vs 32% in the supplemental arm) than infants with plus disease (52% progression in conventional vs 57% in supplemental). Pneumonia and/or exacerbations of chronic lung disease occurred in more infants in the supplemental arm (8.5% conventional vs 13.2% supplemental). Also, at 50 weeks of postmenstrual age, fewer conventional than supplemental infants remained hospitalized (6.8% vs 12.7%), on oxygen (37.0% vs 46.8%), and on diuretics (24.4% vs 35.8%). Growth and developmental milestones did not differ between the 2 arms. CONCLUSIONS: Use of supplemental oxygen at pulse oximetry saturations of 96% to 99% did not cause additional progression of prethreshold ROP but also did not significantly reduce the number of infants requiring peripheral ablative surgery. A subgroup analysis suggested a benefit of supplemental oxygen among infants who have prethreshold ROP without plus disease, but this finding requires additional study. Supplemental oxygen increased the risk of adverse pulmonary events including pneumonia and/or exacerbations of chronic lung disease and the need for oxygen, diuretics, and hospitalization at 3 months of corrected age. Although the relative risk/benefit of supplemental oxygen for each infant must be individually considered, clinicians need no longer be concerned that supplemental oxygen, as used in this study, will exacerbate active prethreshold ROP.


OBJECTIVE: To present ophthalmological outcome data at 5(1/2) years after full term from a natural history cohort of infants who had a birth weight less than 1251 g and were enrolled at 5 centers of the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity (ROP), including eyes without ROP and with a full range of ROP severity. DESIGN: Of the 1199 surviving children in the cohort, 1068 (89.1%) were examined. Study-certified ophthalmologists assessed ROP residua and conducted cycloplegic refractions. Visual acuity was measured by study-trained testers using the Early Treatment for Diabetic Retinopathy Study charts. Eyes that had developed ROP were categorized by the lowest (most posterior) zone and highest (most severe) stage reached during the acute phase of the disease. No eyes that received cryotherapy are included; data analysis included one untreated eye per patient. Fundus outcomes were classified as “unfavorable” if there was macular compromise by retinal folding (more severe than ectopia) or stage 4B or 5 retinal detachment. Visual acuity outcomes of 20/200 or worse were classified as unfavorable. RESULTS: Unfavorable fundus structural outcome occurred in 33 (3.1%) of the 1068 eyes; all 33 eyes had a history of severe ROP. Specifically, unfavorable fundus structure occurred in 62.5% (10/16) of eyes with zone I ROP and in 44.2% (23/52) of eyes with zone II ROP, stage 3+ disease involving more than six 30 degrees -sectors. There were no unfavorable fundus outcomes among eyes that had fewer than 7 clock-hours of stage 3+ ROP in zone II in this cohort. Snellen visual acuity was tested in 1059 eyes, and 5.1% were unfavorable at 20/200 or worse; these unfavorable outcomes were correlated with more severe ROP. In eyes that had zone I ROP, 68.8% (11/16) had unfavorable acuity, and for eyes that had zone II ROP, 7.5% (36/481) had unfavorable acuity results. For eyes with ROP observed only in zone III, 1.8% (2/110) had unfavorable acuity of 20/200 or worse. CONCLUSIONS: Premature infants with birth weights less than 1251 g seldom have poor structural and functional outcomes (3.1% and 5.1%, respectively). All unfavorable fundus structural outcomes and nearly all unfavorable acuity outcomes occurred in eyes with zone I ROP or zone II ROP involving more than 6 sectors of stage 3+ disease.


A case of cortical blindness is presented which developed during H. influenzae type B meningitis. Cortical blindness may appear late in the course of bacterial meningitis, during a period of clinical improvement, and recovery can be expected in 50 per cent of cases. In contrast, when the cause is ischaemic the onset of blindness is immediate and usually recovery is complete. Radiological evidence is presented for the pathological process in meningitis.


Tissue from ten eyes with infantile glaucoma and from 40 normal eyes of fetuses and infants without glaucoma were examined by light and electron microscopy. In normal development, the corneoscleral coat grows faster than the uveal tract during the last trimester, leading to a posterior migration of the ciliary body attachment from Schwalbe's line (5th month) to the scleral spur (9th month), and then to a location behind the scleral spur (postnatally). In infantile glaucoma, the insertion of the anterior ciliary body and iris overlaps the trabecular meshwork, similar to the late fetal position. The trabecular sheets are perforated, and there is no membrane over the surface of the trabecular meshwork. The trabecular beams are thicker than in normal infant eyes. There is both histologic and clinical evidence of traction on the iris root exerted by the thickened trabecular beams. These findings suggest that in congenital glaucoma the thickened beams had prevented the normal posterior migration of the ciliary body and iris root. This traction may compact the thickened trabecular beams, obstructing aqueous humor outflow. Release of the traction by an incision (goniotomy or trabeculotomy) of the thickened meshwork may relieve the obstruction. Of uncertain pathological significance is that there are no vacuoles in the endothelium of Schlemm's canal and there is a broad layer of collagen and amorphous material in the juxtacanalicular connective tissue. The ciliary processes are elongated inward, as if they were pulled by zonular traction (perhaps created by an enlarging diameter of the limbus with a fixed lens diameter).


We studied the change in eye position under general anesthesia in 317 patients undergoing strabismus surgery. We used the prism cover test to measure preoperative eye position, and the prism reflex test (Krimsky's method) to measure the eye position under general anesthesia. Almost all patients had divergence under anesthesia. We calculated a linear equation and curve to relate the eye position under anesthesia to the preoperative eye position. Patients with a normal amount of divergence (within one standard deviation of the mean) had a higher rate of successful surgical results than those with an abnormal amount of divergence under anesthesia (greater than one standard deviation from the mean).


This paper introduces new principles for the design and use of letter charts for the measurement of visual acuity. It is advocated that the test task should be essentially the same at each size level on the chart. Such standardization of the test task requires the use of letters of equal legibility, the same number of letters on each row, and uniform between-letter and between-row spacing. It is also advocated that, combined with the test task standardization, there should be a logarithmic progression of letter size. Charts incorporating these design features have been made. These charts facilitate the use of nonstandard testing distances which might be used when there is low visual acuity, when examining very young or newborns, or when testing at the standard distance, or when it is necessary to validate visual acuity scores or detect malingering. Adjusting the visual acuity score according to the chosen testing distance is simplified by the use of logarithmic scaling.


A reasonable amount of information concerning early refraction and accommodation has been accumulated. The measurements of refraction appear to be reliable and concurrently valid, although more attention should be devoted to both of those issues. There is also some evidence that the measurements can be developmentally significant since adults with histories of early spherical or cylindrical errors exhibit nonreversible visual deficits. The retrospective findings [11, 43] that evidenced this point do not necessarily imply that large refractive errors during infancy invariably yield visual deficits later on. To determine the significance of early errors, their predictive validity must be better estimated. Some evidence indicates that neonatal refractions predict refractions later in infancy. But the actual magnitude of the relationship between early and school aged or adult refractions is uncertain. Longitudinal studies of refraction from birth to at least the elementary school years would yield appropriate data. It is hoped that with the increasing interest in early visual function and the development of accurate, yet rapid techniques of measurement (such as photorefraction), such information will soon be forthcoming. It will be of special interest to unveil the mechanisms that control the development of refraction, in particular, to reveal the developmental relationship between refraction and accommodation.


OBJECTIVE. To compare the efficacy of commonly used forms of eye prophylaxis for newborns with no prophylaxis in the prevention of nongonococcal conjunctivitis. DESIGN. Randomized doubly masked clinical trial. SETTING. University of Washington Hospital and affiliated clinics, Seattle, between 1985 and 1990. SUBJECTS. The medical records of 8499 women were evaluated for possible participation; 2577 were eligible. Of the 758 enrolled, the infants of 630 were evaluable. INTERVENTION. Comparison of silver nitrate, erythromycin, and no eye prophylaxis given at birth for the
prevention of conjunctivitis. MAIN OUTCOME MEASURES. Conjunctivitis during the first 60 days of life and nasolacrimal duct patency in the first 2 days of life. RESULTS. The frequency of impatent tear ducts at the 30- to 48-hour examination did not differ significantly by prophylaxis group. Among the 630 infants randomized and observed, 199 (17%) developed mild conjunctivitis. Sixty-three (63%) of the cases appeared during the first 2 weeks of life. After 2 months of observation, infants allocated to silver nitrate eye prophylaxis at birth had a 39% lower rate of conjunctivitis (hazard ratio = 0.61, 95% confidence interval = 0.39 to 0.97), and those allocated to erythromycin had a 31% lower rate of conjunctivitis (hazard ratio = 0.69, 95% confidence interval = 0.44 to 1.07), than did those allocated to no prophylaxis. CONCLUSION. Silver nitrate eye prophylaxis caused no sustained deleterious effects and even provided some benefit to infants born to women without Neisseria gonorrhoeae. However, the effect was modest and against microorganisms of low virulence. The results suggest that parental choice of a prophylaxis agent including no prophylaxis is reasonable for women receiving prenatal care and who are screened for sexually transmitted diseases during pregnancy.


Chlamydia trachomatis is the most common pathogen associated with conjunctivitis during early infancy in the United States. During a 13-month interval at our medical center 4834 infants were born, 311 of whom (6.4%) had conjunctival specimens tested for chlamydial antigen before the age of 12 weeks. In 44 (14%) of all tested infants, 0.9% of live births) chlamydial antigen was present. Because the rate of asymptomatic maternal chlamydial endocervical colonization is estimated to be 26% at our institution (previous prospective study), we calculated a minimal failure rate for erythromycin ocular prophylaxis of from 7 to 19.5%. A subsequent case-control study revealed that mothers of infants with chlamydial conjunctivitis were more likely to be primiparous (P = 0.03) and experience longer duration of rupture of membranes before delivery (P = 0.006). We conclude that a substantial percentage of infants exposed to Chlamydia develop chlamydial conjunctivitis despite receiving erythromycin ocular prophylaxis.


OBJECTIVES: To present the 3- and 12-month strabismus data from 3030 premature infants with birth weights less than 1251 g enrolled in the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity. DESIGN: Data from the 3- and 12-month examinations conducted at 23 regional study centers were tabulated for all infants. The main outcome measure, ocular motility, was compared with baseline demographic variables and retinopathy of prematurity severity for the worse eye. Findings at 3 months were compared with the incidence of strabismus at 12 months. RESULTS: At 3 months, 200 (6.6%) of the 3030 infants were strabismic. In the 2449 infants examined at both time points, 289 (11.8%) were found to have strabismus at 12 months. Retinopathy of prematurity was significant for strabismus at both 3 and 12 months (P<.001). The presence of strabismus at 3 months was found to be a highly significant predictor of strabismus at 12 months. Anisometropia, abnormal fixation, and unfavorable retinal structure also were significant predictors of
strabismus at 1 year. The total prevalence of strabismus in the first year of life was 14.7%. CONCLUSION: The presence of acute-phase retinopathy of prematurity places the premature infant at increased risk for strabismus.


High-resolution magnetic resonance images of the intracranial optic nerves and chiasm were obtained in 15 patients with severe optic nerve hypoplasia. These were compared, in a double-blind manner, with similar images from 30 age-matched controls. On both coronal and sagittal images, hypoplastic optic nerves were thin and demonstrated signal attenuation when compared with normal optic nerves. All patients with severe bilateral optic nerve hypoplasia also had diffuse chiasmal hypoplasia, which was seen best on coronal images. Patients with unilateral or asymmetrical optic nerve hypoplasia had variable chiasmal abnormalities. The degree to which the magnetic resonance diagnosis of optic nerve hypoplasia matched the clinical diagnosis was highly significant (P less than .001, Fisher's Exact Test) for both coronal and sagittal views of the intracranial optic nerves. Oblique axial and coronal views of the orbital optic nerves did not reliably distinguish optic nerve hypoplasia from normal optic nerves. High-resolution magnetic resonance imaging is a useful diagnostic modality to identify small optic nerves neuroradiologically.


In the Ndongue Hospital in Cameroon 449 new-born babies were examined for a month to check on the occurrence of ophthalmia neonatorum. Silver nitrate eyedrops 1% were applied at birth. The incidence of ophthalmia neonatorum was 19.4%; Chlamydia was found in 8 cases (incidence 1.8%) and gonococci in 4 cases (0.9%). Chemical conjunctivitis was suspected in 13 cases (2.9%). Slight conjunctivitis was seen in 68 cases and severe conjunctivitis in 19 cases (4.2%). A sexually transmitted agent was demonstrated in 6% of the slight cases and in 42% of the severe cases. Ophthalmia neonatorum due to Chlamydia was significantly more severe and without specific features than those due to other agents. Although silver nitrate is still considered to be one of the most effective prophylactic measures for gonococcal ophthalmia neonatorum, in 4 cases this disease still developed. Silver nitrate is ineffective against Chlamydia and is also often the cause of a chemical conjunctivitis. The search is advocated for more efficient medicines in Africa which are effective against both Chlamydia and resistant gonococci.


A hypothesis, previously proposed, of tight medial rectus muscles in conjunction with tight lateral rectus muscles associated with exodeviations as a cause of lateral incomitancy in intermittent exotropia is supported by clinical management. Three patients with these findings underwent bilateral medial rectus and lateral rectus recessions by means of the adjustable rectus recession technique. Primary position alignment was achieved, and rotations were balanced with the alleviation of the lateral incomitancy. The lateral rectus muscles were recessed an amount more than usual in order to compensate for the recession of the medial rectus muscles.


From November, 1989, to October, 1991, 4544 neonates were born at our hospital. Neonatal ocular prophylaxis immediately after birth was used with 1% tetracycline ophthalmic ointment in 1156 neonates, 0.5% erythromycin ophthalmic ointment in 1163 neonates and 1% silver nitrate drops in 1082 neonates. No prophylaxis for neonatal conjunctivitis was given to 1143 neonates. A total of 302 infants (6.7%) developed conjunctivitis during the first 4 weeks of life. Between December, 1991, and January, 1992, 425 neonates were born at our hospital and all were given 0.5% erythromycin ophthalmic ointment twice in the first 24 hours after birth for ocular prophylaxis. Thirty-one (7.3%) infants developed conjunctivitis during the neonatal period. The incidence rates of neonatal chlamydial conjunctivitis in the tetracycline, erythromycin, silver nitrate, no prophylaxis and erythromycin twice groups were 1.3, 1.5, 1.7, 1.6 and 1.4%, respectively. We conclude that neonatal ocular prophylaxis with erythromycin (one or two doses) or tetracycline or silver nitrate does not significantly reduce the incidence of neonatal chlamydial conjunctivitis compared with that in those given no prophylaxis.


OBJECTIVE: To describe short-term structural outcomes and associated ocular complications in premature infants treated with diode laser ablation for retinopathy of prematurity. METHODS: The records of all infants who were diagnosed as having threshold retinopathy of prematurity and treated with diode laser therapy at our hospital from January 1, 1992, through December 31, 1996, were retrospectively reviewed. Sixty-four eyes reached threshold during this period. Three eyes received cryotherapy in addition to laser treatment and were excluded, leaving 61 eyes eligible for review.
RESULTS: Of the 61 eyes with threshold disease treated exclusively with diode laser, 4 (7%) had zone I disease and 57 (93%) had zone II disease at the time of initial laser treatment. Three (5%) of the 61 eyes progressed to stage 4 disease (2 eyes, stage 4A; 1 eye, stage 4B). There were no cataracts or other ocular complications noted secondary to laser treatment based on short-term follow-up (mean follow-up, 120 days). CONCLUSION: In this population of infants, diode laser ablation appears to be a safe and effective treatment for threshold retinopathy of prematurity.


Primary infantile glaucoma, commonly termed congenital glaucoma or trabeculodygenesis, is an unusual, inherited connal anomaly of the trabecular meshwork and anterior chamber angle which leads to obstruction of aqueous outflow, increased intraocular pressure, and optic nerve damage. Its pathogenesis is still disputed; most observers have not been able to document ultrastructurally a continuous endothelial membrane, as initially advanced by Barkan. Medical therapy for primary infantile glaucoma is accorded a supportive role; the primary, definitive treatment is surgical. Both goniotomy and trabeculotomy ab externo give similarly good results in the majority of patients. The prognosis in this disease is related to the time of its initial presentation, initial surgical intervention, degree of optic nerve damage, nature and quality of corneal enlargement and astigmatism, progressive refractive error, and anisometropic amblyopia. The inability to easily quantify visual acuity and extent of visual loss in neonates makes these parameters less helpful in following patients than measurement of corneal diameter and intraocular pressure. However, even these data should not be relied upon exclusively to determine the quality or quantity of success in primary infantile glaucoma.


Prospective follow-up studies were done on 62 term infants who were treated as neonates for clinical evidence of postasphyxial encephalopathy. Computed tomographic studies were done during the first two weeks of life and repeated at six months of age. All children were followed a minimum of 18 months, at which time they underwent a psychometric and a neurologic evaluation. Major neurodevelopmental sequelae consisted of: hydrocephalus; spastic quadriplegia, hemiplegia, or diplegia; or a mean Bayley score less than 70. Major sequelae were present in 29 (47%) of the children: all were severely handicapped. Five other children scored between 70 and 85 on the Bayley test. Computed tomographic scans were highly predictive of status at 18 months. Eleven of the 15 with intraventricular or parenchymal hemorrhage were severely handicapped. Eighteen of 20 with extensive areas of hypodensity of the white and gray matter (neonatal CT) were abnormal at 18 months. All but two were severely handicapped. The results suggest that CT studies are very useful in the care of the asphyxiated term infant who has clinical signs of encephalopathy.


BACKGROUND. Retinopathy of prematurity is a disease affecting the blood vessels of the retina in premature infants that may result in scarring, retinal detachment, and loss of vision. An association between this condition and the exposure of premature infants to supplemental oxygen has been postulated, but the relation between retinopathy of prematurity and blood oxygen levels has not been defined. The purpose of this study of a cohort of preterm infants was to correlate the incidence and severity of retinopathy of prematurity with the duration of exposure to different ranges of oxygen tension as measured by transcutaneous monitoring (tcPO2). METHODS. One hundred one premature infants (birth weight, 500 to 1300 g) requiring supplemental oxygen had continuous monitoring of tcPO2. The number of hours during which the tcPO2 was 80 mm Hg or higher was tabulated for each infant during the first four weeks of life. RESULTS. There was a significant association between the amount of time that the tcPO2 was greater than or equal to 80 mm Hg and the incidence and severity of retinopathy of prematurity. The odds ratio for each 12-hour period in which the tcPO2 was 80 mm Hg or higher was tabulated for each infant during the first four weeks of life. CONCLUSIONS. This study supports an association between the incidence and severity of retinopathy of prematurity and the duration of exposure to arterial oxygen levels of 80 mm Hg or higher, measured transcutaneously.
The changes occurring in retrolental fibroplasia were studied in 97 premature infants with fundus photography and
fluorescein angiography. The macrovascular lesion of proliferative retrolental fibroplasia was identified as a functioning
arteriovenous shunt. The microvascular abnormalities were capillary tufts, collaterals, capillary-free zones, and
neovascular membranes. Regression was signaled by the ingrowth of capillaries from the shunt into the avascular retina.
Cicatrization was characterized by persistence of the vascular abnormalities of the proliferative phase and organization of
the avascular retina into a contracting scar.


Optic nerve hypoplasia is a non-progressive condition characterised by subnormal vision and a subnormal number of
optic nerve axons. It may be unilateral or bilateral, isolated or combined with other defects. Analysis of fundus
photographs from a series of 7 patients with a stationary abnormality of different degrees showed that the functional
defects could be closely correlated with defects in the retinal nerve fibre layer. Our observations show that the condition
has a wide range of both functional and anatomical defects and that a subnormal diameter of the optic disc is not a
requisite for the diagnosis. Presumably, there is also a wide variety of causes, not only a primary failure of development
of retinal ganglion cells. We suggest that optic nerve hypoplasia can be viewed as a non-specific manifestation of damage
to the visual system, sustained any time before its full development.

The incidence and aetiology of ophthalmia neonatorum have been estimated over a 7-month period in Franceville, a
semi-rural community in south-eastern Gabon. Chlamydia trachomatis was the most frequently observed pathogen, being
isolated from 17 babies (2.7% of births), and Neisseria gonorrhoeae was recovered from 12 (1.6% of births). 5 of 17
cases of chlamydial conjunctivitis were in infants less than 5 d old as opposed to 9 in the typical 5 to 10-days-old group.
As expected, most cases of gonococcal ophthalmia neonatorum occurred in the first 5 d of life with cases in older infants
often not accompanied by a granulocytic response. Chlamydial conjunctivitis was usually unilateral whereas other cases
were most frequently bilateral.

structural and functional outcome. Multicenter Trial of Cryotherapy for Retinopathy of Prematurity Cooperative Group." Arch
Ophthalmol 114(9): 1085-1091.
OBJECTIVE: To investigate the structural and functional outcome at age 4 1/2 years of eyes that had partial retinal
detachment (RD) at 3 months after the occurrence of threshold retinopathy of prematurity; these eyes were involved in the
Multicenter Trial of Cryotherapy for Retinopathy of Prematurity study. METHODS: Of the 531 eyes in the randomized
portion of the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity study, 61 had partial RD at the 3-month
examination. The extent of retinal involvement was recorded, and the presence of a foveal detachment was noted. At the
examination conducted at age 4 1/2 years, the fundus structure was graded into cicatricial retinopathy of prematurity
outcome categories by study ophthalmologists, and the visual acuity was measured by masked examiners using the
Teller Acuity Card procedure and the crowded HOTV recognition acuity test. RESULTS: Of the 61 eyes, 7 eyes continued
to have partial RD at age 4 1/2 years. Of the remaining eyes, 20 eyes had structural outcomes classified as favorable in the
Multicenter Trial of Cryotherapy for Retinopathy of Prematurity study, and 27 eyes had unfavorable structural
outcomes. Data were unrecordable for 3 eyes and missing for 4 eyes. No difference in outcome was found for eyes with
partial RD at 3 months that had undergone cryotherapy vs eyes that had served as controls. Only 6 eyes had a visual
acuity better than 20/200. When partial RDs did not involve the fovea at 3 months, structural and functional outcomes at
4 1/2 years were better than when RDs involved the fovea. The best predictor of outcome was the extent of RD at 3
months. CONCLUSIONS: Partial RD present 3 months after threshold retinopathy of prematurity is unstable anatomically,
and the visual outcome is generally poor. Structure and function at 4 1/2 years are related to the extent of RD and the
involvement of the fovea at 3 months.


Noncycloplegic refractions of 1,000 children aged 0-6 years revealed a high incidence of astigmatism, especially in the
first 2 years of life. Before age 41/2 years, most of the astigmatism was against-the-rule and after that age most was
with-the-rule. Of 19 children who did not show astigmatism in the first year, only one acquired it by 4 years. Of 29
children who had large amounts of astigmatism in the first year, all showed elimination or a large reduction in the
amount of the cylindrical error by 4 years. These results are relevant to the etiology of astigmatism.


Opinions differ concerning the efficacy of prophylaxis against neonatal chlamydial and gonococcal conjunctivitis. From January 1986 through June 1988, we gave all infants born at Kings County Hospital Medical Center one of three prophylactic agents -- silver nitrate drops, erythromycin ophthalmic ointment, or tetracycline ophthalmic ointment. The treatments were rotated monthly. Gonococcal ophthalmia occurred in 8 of the 12,431 infants born during the study (0.06 percent), 1 in the silver nitrate group, 4 in the erythromycin group, and 3 in the tetracycline group (P not significant). Seven of these infants were born to women who had received no prenatal care. From September 1985 through December 1987, we screened 4357 pregnant women for cervical chlamydial infection, of whom 341 (8 percent) had positive cultures. Of their offspring, 230 were evaluated for neonatal chlamydial conjunctivitis; the incidence was 20 percent in the silver nitrate group, 14 percent in the erythromycin group, and 11 percent in the tetracycline group (P not significant). We conclude that neonatal ocular prophylaxis with either erythromycin or tetracycline ophthalmic ointment does not significantly reduce the incidence of chlamydial conjunctivitis in the offspring of mothers with chlamydial infection as compared with silver nitrate, and that better management of maternal chlamydial infection is therefore required. We also conclude that there is a small but appreciable incidence of neonatal gonococcal ophthalmia that could be prevented by better prenatal screening and treatment of maternal gonococcal infection.


To study the long-range results of surgically treated intermittent exotropia, 100 consecutive patients have been followed up for an average of 6.1 years. In all cases, the initial procedure was bilateral recession of the lateral rectus muscles. The overall functional cure rate was 78%. To accomplish this result, 27 patients were operated on a second time, 21 for undercorrection and six for overcorrection. A number of patients cooperated very poorly or were lost to follow-up while still under treatment. Had these patients been eliminated from the series, the cure rate would have been greater than 90%. In this study, bilateral recession of the lateral rectus muscles corrected the distant measurement more than the near measurement only with the divergence excess type of deviation. Also, this procedure was not notably more effective with divergence excess than with basic-type intermittent exotropia.


Iris circulation was investigated by fluorescein iris angiography in 33 human eyes before and after tenotomy of one or more recti for correction of strabismus. Tenotomy of one or both horizontal recti produced no appreciable circulatory disturbance in the iris, but tenotomy of superior or inferior rectus produced circulatory delay in superior temporal or inferior temporal sectors, respectively, of the iris. When tenotomies of a horizontal and one or two vertical recti were combined, the defect occurred in the region of the vertical rectus only. Blood supply of the nasal half of the iris was usually not disturbed by tenotomy of the vertical and/or medial rectus. The findings indicate that the blood supply of the iris is segmental and suggest that, in strabismus surgery, cutting the two vertical recti along with the lateral rectus may subject the eye to the risk of anterior segment ischemia. In the light of the present findings, physiological anatomy of the blood supply of the iris and role of the major arterial circle of the iris are discussed.


We investigated an outbreak of erythromycin-resistant *Staphylococcus aureus* conjunctivitis in a hospital newborn nursery that used erythromycin eye ointment to prevent ophthalmia neonatorum. Cases occurred in 2 clusters; 20 (14%) of 146 infants in the nursery developed conjunctivitis from July through October, 1987; and 5 (7%) of 69 infants in the nursery developed conjunctivitis during April and May, 1988. A case-control study of the first cluster demonstrated that culture-confirmed cases were more likely than controls to have received prophylactic erythromycin eye ointment or their initial bath from one nurse (odds ratio, 9.0; P = 0.01) or to have been delivered by one physician (odds ratio, 12.7; P = 0.03). The nurse was the only staff person to have a nasopharyngeal culture which yielded erythromycin-resistant S. aureus. Control measures, instituted in October, 1987, included using silver nitrate drops instead of erythromycin eye ointment for prophylaxis; however, in January, 1988, the hospital resumed use of erythromycin eye ointment. No additional cases were identified until mid-April, 1988, when the second cluster of cases occurred. At that time the hospital reinstated the use of silver nitrate and no additional cases were identified. This investigation illustrates the potential for conjunctival infection with an antimicrobial-resistant pathogen when antimicrobials are used to prevent ophthalmia neonatorum.


Analysis of the optics of photorefraction showed that, for short camera-to-subject distances, the function relating image size to defocus of the eye is not symmetrical for errors of focus in front of and behind the camera. This asymmetry is exploited in the new method of isotropic photorefraction, in which the supplementary cylinder lenses of the original orthogonal photorefractors are replaced by defocusing of the camera lens itself. By comparing photographs taken with the camera focused in front of and behind the subject, the sign of the eyes' defocus (myopic or hyperopic relative to the camera) can be determined. The axis of any astigmatism is readily apparent as the direction in which the photorefractive images are elongated. The method is well adapted for the refractive screening of infants and young children.


In the largest study to date of bacterial flora in newborns, we cultured the conjunctivae of 100 infants within 15 minutes after vaginal delivery and before any antimicrobial agents had been applied to the eye. All cultures were intensively analyzed for anaerobic and aerobic bacteria. By far the largest group of bacteria isolated were microaerophilic, such as Lactobacillus species and diptheroids, accounting for 46.8% of positive cultures and 62.3% of all bacteria isolated. The second largest group were true anaerobic bacteria, such as Bacteroides and Propionibacterium species. The smallest group were aerobic bacteria. This incidence of non-aerobic bacteria in the conjunctiva of newborns is the highest reported to date. Our finding should alert clinicians to consider non-aerobic, especially microaerophilic, bacteria in the differential diagnosis of ophthalmia neonatorum. The high rate of supposedly sterile cultures reported in other studies may be explained at least partially by improper isolation of non-aerobic bacteria.


PURPOSE: The agents currently used to prevent ophthalmia neonatorum are less than optimal, with reports indicating evidence of bacterial resistance, ineffectiveness, and toxicity. Povidone-iodine ophthalmic solution, which has been shown to be effective in the preoperative preparation of the eye, generates no resistance, is an effective antimicrobial agent, and has low toxicity. We evaluated the effectiveness and safety of povidone-iodine for ophthalmia neonatorum prophylaxis.

METHODS: A bacterial culture was taken from the conjunctiva of each eye of 100 infants within 30 minutes of birth. A drop of 2.5% povidone-iodine solution was then placed on one eye, while the other eye received either one drop of silver nitrate 1% ophthalmic solution or 0.5% erythromycin ointment. Conjunctival bacterial cultures were again taken two to four hours after birth. At each culture and at 24 hours after birth, the eyes were examined for toxic changes. To measure the effectiveness of the medications, the number of bacterial colony-forming units and species from each culture was compared. RESULTS: All three agents significantly reduced the number of colony-forming units, but povidone-iodine caused the most significant decrease. The number of species was reduced significantly by povidone-iodine (P = .00051) and silver nitrate (P = .007), with povidone-iodine yielding the most significant decrease. Erythromycin did not significantly reduce the number of species. Silver nitrate demonstrated more ocular toxicity at the 24-hour determination point than did either of the other two medications (P < .001). CONCLUSIONS: Povidone-iodine 2.5% ophthalmic solution is an effective antibacterial agent on the conjunctiva of newborns and causes less toxicity than silver nitrate.

OBJECTIVE: To evaluate optic disc morphologic features in children with periventricular leukomalacia (PVL). PATIENTS AND METHODS: Seventeen children with PVL (patient group) were compared with 17 sex- and age-matched, full-term healthy control children (control group). Clinical ophthalmological examination and digital image analysis of fundus photographs were performed in all children. In children with PVL, cerebral imaging was performed. RESULTS: Children with PVL had larger optic disc cupping (P<.02) than did control children. A large proportion of the children with PVL had strabismus, nystagmus, and restricted visual fields. CONCLUSION: Our study indicates that optic nerve hypoplasia in children with PVL is often associated with a normal-sized optic disc with a large cup. This unusual form of optic nerve hypoplasia most likely results from transsynaptic degeneration of optic axons caused by the primary bilateral lesion in the optic radiation.


Two visually impaired children with occipital infarctions are presented. One patient has profound impairment of his primary visual pathway but has good vision for traveling, while the other child presented with the symptoms in reverse. We believe that these two patients provide further evidence that the primary visual pathway is used for conscious visual analysis and that the collicular visual system serves as the subconscious visual guidance for locomotion.


BACKGROUND. Retinopathy of prematurity (ROP) is the leading cause of blindness among premature infants. A recent National Eye Institute-sponsored prospective, multicenter trial investigating the use of cryotherapy for treatment of ROP demonstrates a significant reduction in blindness and low vision for patients with sight-threatening (stage 3+) ROP. METHODS: A microsimulation model is presented to determine the cost-effectiveness of cryotherapy for ROP. Simulations are performed for three subpopulations of premature infants with birth weights 500 through 749 g, 750 through 999 g, and 1000 through 1249 g, and for three screening strategies—weekly, biweekly, and monthly. RESULTS. Appropriately timed screening for and treatment of ROP is predicted to result in a gain of 3899 to 4648 quality-adjusted-life-years and a net governmental budgetary savings of $38.3 to $64.9 million for each annual US birth cohort of 28,321 prematurity infants (500 through 1249 g). The cost per quality-adjusted-life-year gained is $2488 to $6045, depending on different screening strategies. CONCLUSIONS. Of greatest importance is the finding that properly timed screening and treatment for ROP is not only cost saving but may save approximately 320 infants per year from a lifetime of blindness.


The relation between PaO2 and retrolental fibroplasia (RLF) was studied prospectively in 719 premature infants born in or treated in the intensive care units of a group of university hospitals. Blood gas studies were performed on 589 of these infants, 66 of whom had a diagnosis of RLF; in 27 of these 66, some grade of mostly nonblinding cicatricial disease developed. The frequency of RLF was highest among infants of lowest birth weight. A multivariate statistical method was used to analyze simultaneously the effect of possible etiologic factors associated with RLF. The occurrence of RLF was found to be unrelated to PaO2, as determined by the limited information available from intermittent sampling. RLF is associated with concentration of oxygen administered in the lightest birth weight group, but the strongest association, aside from birth weight, was with time in oxygen. None of the other variables involving blood chemical values appeared...
to be associated with RLF. The severity of cicatricial RLF is clearly greater in infants weighing less than 1,200 g at birth. Conservative administration of oxygen may have been responsible for failure to demonstrate quantitative association between PaO2 levels and disease. Agreement between the observed and predicted numbers of infants with RLF demonstrate the strength of the multivariate technique employed in making the statistical analyses.


A laser optometer and an auditory feedback system were used to assess the accommodative response in normal eyes, and in the dominant and amblyopic eyes of amblyopes. Although the accommodative response improved in amblyopic eyes when these eyes achieved steady foveal fixation it is not clear how much of the subnormal acuity in amblyopic eyes is due to eccentric fixation, unsteady fixation, inappropriate accommodation, refractive error or some combination of these factors.


The authors conducted this study to determine the etiologic agents of conjunctivitis in early infancy. From 1985 to 1990, 630 infants enrolled in a randomized, controlled, double-masked study of eye prophylaxis were observed for 60 days after delivery for signs of conjunctivitis. The following isolates were categorized as pathogens: Haemophilus influenzae, Streptococcus pneumoniae, Neisseria cinerea, Klebsiella pneumoniae, and Chlamydia trachomatis. Using conditional logistic regression for analysis of 97 infant pairs, the authors identified isolates categorized as pathogens almost exclusively among cases (odds ratio (OR) = 18.0, 95% confidence interval (CI) 2.3-128). Among the microorganisms which have not usually been regarded as pathogens in the etiology of infant conjunctivitis, Streptococcus mitis was the only microorganism associated with an increased risk of conjunctivitis (OR = 5.3, 95% CI 1.8-15.0). The findings concerning the species of bacteria most often associated with conjunctivitis, as well as the finding that method of delivery is unimportant, suggest that bacteria were transmitted to the infants' eyes after birth and not from the birth canal.


A series of 38 patients with regressed retinopathy of prematurity were compared with control groups of babies having similar gestational age who did not have retinopathy of prematurity, and to a control group of full-term babies. A substantially higher incidence of strabismus, amblyopia, and high refractive errors was found in the group with retinopathy of prematurity as compared with both other groups. In general, these patients had a successful response to standard amblyopia and strabismus management. No substantial difference was found between the refractive errors of the babies in the premature control group and the term control group.


Seven patients with structural abnormalities of one or both of their optic nerves had decreased visual acuity partly due to functional amblyopia. They were treated with conventional amblyopia therapy and their visual acuities improved. Functional amblyopia can coexist either secondary to, or coincidentally with, structural optic nerve abnormalities.


From a public health point of view gonococcal ophthalmia neonatorum (GON) is important as it can rapidly lead to blindness. The frequency of GON is determined by the prevalence of maternal gonococcal infection. In most industrialized countries the prevalence of gonorrhea in pregnant women is less than 1%; in developing countries the rates are between 3% and 15%, more than 50% being due to penicillinase-producing Neisseria gonorrhoeae strains (PPNG). The rate of transmission from mother to newborn is between 30% and 50%. Strategies for the control of GON include: (1) prevention of gonococcal infection in women of childbearing age, (2) detection and treatment of gonococcal infection in pregnant women, (3) eye prophylaxis in the newborn at birth, and (4) diagnosis and treatment of GON. Eye prophylaxis by the instillation immediately after birth of either 1% silver nitrate eye drops or 1% tetracycline eye ointment is very effective. This reduces the GON incidence by 80% to 95% and is highly cost-effective, particularly in high-risk settings.

PIP: Ophthalmia neonatorum is a purulent conjunctivitis in infants less than 30 days old, where the Gram stain of an eye smear shows at least one polymorphonuclear leukocyte per high-power field. The two main causes of the condition are Neisseria
gonorrhea and Chlamydia trachomatis. Gonococcal ophthalmia neonatorum (GCON), however, tends to appear earlier and to be more severe than chlamydial infection. GCON can rapidly lead to blindness. The frequency of GCON is determined by the prevalence of maternal gonococcal infection. In most industrialized countries, the prevalence of gonorrhea in pregnant women is less than 1%. In developing countries, however, prevalence is in the range of 3-15%, with more than 50% of cases caused by penicillinase-producing Neisseria gonorrhoea. A total of 136 injections was given to 83 patients for strabismus (99 injections), blepharospasm (29 injections), and spastic entropion (eight injections). All four patients with entropion experienced temporary benefits and early recurrence; one injection resulted in temporary paralytic ectropion. Two of 13 patients treated for blepharospasm developed transient


In a Nairobi hospital where ocular prophylaxis against ophthalmia neonatorum has been discontinued, 1,019 women were screened for Neisseria gonorrhoea and Chlamydia trachomatis during labour and 7 and 28 days postpartum. The prevalence of gonococcal infection was 7% and that of chlamydia was 29%, 52.4% of gonococcal isolates produced penicillinase. The incidence of ophthalmia neonatorum was 23.2 per 100 live births, and incidences of gonococcal and chlamydial ophthalmia were 3.6 and 8.1 per 100 live births, respectively. Of 181 cases of neonatal conjunctivitis, 31% were caused by C trachomatis, 12% by N gonorrhoea, and 3% by both. In 67 babies exposed to maternal gonococcal infection and 201 exposed to maternal chlamydial infection, rates of transmission to the eye were 42% and 31%, respectively, and to the throat were 7% and 2%. Gonococcal transmission rate was higher in mothers with concomitant chlamydial infection (68%; p = 0.01). Postpartum endometritis was associated with ophthalmia neonatorum (p less than 0.001). Ocular prophylaxis at birth for gonococcal ophthalmia should be reintroduced.


We reviewed the clinical courses and computed tomographic (CT) and magnetic resonance imaging (MRI) scans of 30 infants and children with cortical blindness following hypoxic insults. The degree of injury to the striate and parastriate cortices and the area of the optic radiations were graded from 0 to 4 by a neuroradiologist. Only two children had normal scans of the posterior visual pathway and both had favorable visual outcomes. The visual recovery differed significantly with respect to the age at which the hypoxic insult occurred and CT and MRI abnormalities in the area of the optic radiations, but not with abnormalities in the striate or parastriate cortices. Our results suggest that CT and MRI scanning are helpful in prognosticating the visual potential of children with hypoxic cortical blindness.


Optic nerve hypoplasia is an easily overlooked, nonprogressive developmental anomaly which results in a wide range of visual deficits. It is frequently associated with clinically significant central nervous system and endocrine abnormalities. Maternal substance abuse is increasingly recognized in many cases. A supranormal regression of optic nerve axons in utero, rather than a primary failure of differentiation, is proposed as the pathogenesis.


We conducted a retrospective study of 55 cases to investigate the effectiveness of three different surgical procedures (medial rectus muscle recession and lateral rectus muscle resection in 16 cases, the Hummelsheim procedure in 27 cases, and the Jensen procedure in 12 cases) in the treatment of lateral rectus muscle paralysis. All three procedures were equally effective. Shorter duration of lateral rectus muscle paralysis, greater preoperative lateral version, less contracture of the medial rectus muscle, and a traumatic origin for the sixth cranial nerve damage had a significant effect (P less than .05) on the final results of surgery.


The dark focus of accommodation for an individual observer correlates highly with the magnitude of night, empty field, and instrument myopia. These anomalous myopias are interpreted as resulting from the passive return of accommodation to an individually determined intermediate dark focus when the stimulus for accommodation is degraded or absent, or when the need for accommodation is eliminated.


A total of 136 injections was given to 83 patients for strabismus (99 injections), blepharospasm (29 injections), and spastic entropion (eight injections). All four patients with entropion experienced temporary benefits and early recurrence; one injection resulted in temporary paralytic ectropion. Two of 13 patients treated for blepharospasm developed transient
bilateral blepharoptosis. Temporary and related sequelae of extraocular muscle injection included one periocular hemorrhage, one total ophthalmoplegia, and a 44% incidence (29 of 66 patients) of blepharoptosis, which in two patients lasted more than six months. Within three days of injection one patient developed homolateral acute herpes simplex keratitis and a second died of an acute myocardial infarction. No causal relationship for these events has been established.


Fifty-one patients with congenital optic nerve hypoplasia (CONH) were reviewed. It was found that the risk of having an affected child is higher in an adolescent mother, and that maternal alcohol or drug abuse may be important factors. Frequently the disorder is associated with other neuropsychiatric handicaps, and with neuro-endocrine abnormalities. The findings suggest that CONH probably is not a homogeneous group of disorders; some may be caused by primary failure of differentiation of the retinal ganglion cells, while others may be the product of an acquired transsynaptic degeneration of optic-nerve fibres.


Twenty-five children born at term with perinatal asphyxia were studied at age 2.5 to 4.5 years to evaluate visual function and to determine the prognostic value of postnatal assessments of visual outcome. Postnatal assessments included several visual evoked potentials and electoretinograms in the first week of life. Follow-up assessments included flash and pattern visual evoked potentials, visual evoked potential threshold measurements, and clinical eye examinations. Nineteen children had normal visual function, three were visually impaired, and three remained blind. A strong association was found between normal, abnormal, or absent visual evoked potentials in the early postnatal period and long-term visual outcome (P less than .0001). Other perinatal indicators of asphyxia, including neurologic status, Apgar scores, and arterial pH values, were poor predictors of visual outcome. The risk of visual impairment was limited to those survivors with neurodevelopmental deficits.


The majority of patients with monocular limitation of elevation probably have an inferior restriction as the cause of limited elevation. When there is no vertical deviation in the primary position, the absence of a superior rectus palsy is most likely. Treatment of cases with inferior restrictions consists of recession of the tight inferior rectus and conjunctival recession. If the vertical deviation in the primary position is large enough, inferior rectus recession may be combined with superior rectus resection. When superior rectus palsy is the cause of limited upgaze, transposition surgery, utilizing the lateral and medial rectus muscles transposed to the superior rectus insertion, can be utilized. This often results in significant correction of the vertical deviation in primary gaze, but only yields modest improvement of elevation. Restrictions, which may also be present, must first be released before transposition surgery can succeed. The evidence suggests that rarely need the mechanism of "double elevator palsy" be invoked to explain monocular limitation of elevation. When there is true weakness of elevation, superior rectus palsy alone can account for the clinical findings.


Substitution of erythromycin ointment for silver nitrate in the prophylaxis of gonococcal ophthalmia neonatorum (GON) was accompanied by eight infections in 749 (1.1%) well-born and 21 infections in 285 (7.4%) intensive care infants during an eight-month period. This was significantly higher than previous rates of ophthalmitis during the use of silver nitrate, 0.3% (5/1877) and 2.1% (19/904) for well and intensive care infants, (P less than 0.01). Multiple bacteria were isolated, polymicrobial infection occurred frequently and the pattern of bacterial isolates did not favor cross-contamination.
between infants. Observation suggested the frequent unintentional introduction of ungloved fingers into neonatal eyes during attempts to insert erythromycin ointment. Replacement of ophthalmic ointment with a liquid tetracycline preparation resulted in a decrease in non-gonococcal ophthalmits to rates similar to the baseline period 8/997 (0.8%) for both nurseries. The increased number of infections appear related to the ointment vehicle, difficulty in its application and the mechanical introduction of bacteria. It is inferred that appropriate application of liquid medication also reduced the risk of inadequate prophylaxis.


Premorbid ophthalmoscopic and histopathologic findings were correlated in a case of bilateral optic nerve hypoplasia in a 9-month-old infant with bilateral hydranencephaly. The double-ring sign was due to an extension of retina and retinal pigment epithelium (RPE) over the outer portion of the lamina cribrosa. The outer ring was the junction between sclera and lamina cribrosa, and the inner ring was the termination of the RPE. The center of the inner ring was the hypoplastic nerve head, which appeared whitish because of fibroglial tissue surrounding the central retinal vessels where they entered the optic nerve head. We speculate that an in utero vascular insult, after the third month of development, led to cystic cavitation of the anterior cerebral hemispheres, with subsequent retrograde degeneration of developing retinal ganglion cells.


Birth asphyxia is a major cause of neonatal mortality and morbidity. It remains difficult to predict accurately neurologic outcome among survivors, particularly infants with moderate hypoxic-ischemic encephalopathy. Visual evoked potential (VEP) is a reproducible measure of cortical function and reflects acute changes in central nervous system status secondary to asphyxial insult. We performed serial VEPs on 36 term infants with documented birth asphyxia to investigate the relationship between VEPs and neurodevelopmental outcome at 18 months of age. Fourteen infants were neurologically intact at subsequent examination; all had normal VEPs during the first week of life. Twenty-two infants had died or were significantly handicapped at 18 months of age; 20 had abnormal VEPs persisting beyond day 7 of life. Abnormal VEPs accurately predicted abnormal outcome (100%) and were both sensitive (91%) and specific (100%). In 20 infants who were classified as moderately asphyxiated according to the criteria of Sarnat and Sarnat, even greater accuracy, sensitivity, and specificity (all 100%) were observed. VEPs demonstrate good correlation with neurodevelopmental outcome in term infants with birth asphyxia and provide accurate prognostic information useful in the clinical management of these infants.


A newborn with severe ocular herpes simplex virus (HSV) type 2 infection acquired in utero is presented to exemplify problems in diagnosis and management. A review of 297 newborns with HSV type 1 or type 2 infection reveals that about one-fifth demonstrate ocular involvement including one or more of the following: microphthalmia, conjunctivitis, keratitis, chorioretinitis, optic neuritis and cataracts.


In the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity (ROP), 4099 infants weighing less than 1251 g at birth underwent sequential ophthalmic examinations, beginning at age 4 to 6 weeks, to monitor the incidence and course of ROP. Overall, 65.8% of the infants developed ROP to some degree; 81.6% for infants of less than 1000 g birth weight. As expected, ROP incidence and severity were higher in lower birth weight and gestational age categories. Black infants appeared less susceptible to ROP, of all severity categories, than nonblack infants. The timing of retinal vascular events correlated more closely with postconceptional age than with postnatal age, implicating the level of maturity more than postnatal environmental influences in governing the timing of these vascular events. These results include the current incidence of various severity stages of ROP found in the United States and provide new insight into the development of ROP.


Seventeen children, born of severely diabetic mothers, exhibited segmental optic nerve hypoplasia with normal visual acuity and altitudinal or sector field defects corresponding to the hypoplastic areas of the disc. This is the first group of patients with optic nerve hypoplasia in which a possible cause has been identified. Optic nerve hypoplasia is neither a rare abnormality usually associated with serious central nervous system defects, nor is it always accompanied by decreased visual acuity.


What is the cause of glaucoma in Sturge-Weber syndrome? Looking for the answer to this puzzling question, we examined 21 patients with the disease. Sixteen patients had glaucoma: three bilateral and 13 unilateral. Episcleral hemangiomas were visible in all glaucomatous eyes. In general, the more extensive the hemangioma, the more severe was the glaucoma. During gonioscopy, blood could easily be made to reflux into Schlemm's canal of glaucomatous eyes. Often the canal separated into multiple fine channels. Episcleral venous pressure, which we measured in 11 patients, was high in all glaucomatous eyes. These observations suggest that glaucoma in Sturge-Weber syndrome is caused by elevated episcleral venous pressure. Most likely, veins draining aqueous from the canal of Schlemm are part of an intrascleral or episcleral hemangioma. The canal of Schlemm itself may be part of the hemangioma. Arteriovenous shunts in the hemangioma raise episcleral venous pressure, which in turn elevates intraocular pressure.


During the course of the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity (CRYO-ROP), 98 infants (129 eyes) from the randomized segment of the trial developed total retinal detachment from retinopathy of prematurity (ROP) before the 1-year examination. The authors report as a case series the results of acuity card assessment of monocular grating acuity at 1 year in 53 infants (71 eyes) postvitrectomy and in 45 infants (58 eyes) who had retinal detachments but who did not undergo retinal reattachment surgery. The decision to undertake and the surgical technique used for a retinal reattachment procedure was not part of the randomized CRYO-ROP trial. Two eyes of one infant had pattern vision at the lowest measurable threshold after vitrectomy. None of the remaining eyes that had undergone vitrectomy and none of the eyes that did not undergo vitrectomy showed evidence of pattern vision. The relatively poor visual outcomes in this case series suggest that efforts are well-spent in attempting to prevent retinal detachment in ROP.


PURPOSE: The purpose of the study was to examine spherical equivalent refractive errors, especially myopia, at six ages between 3 months and 5 1/2 years post-term in preterm children with birth weights of less than 1251 g. DESIGN: A cohort study. PARTICIPANTS: There were a total of 827 participants in the multicenter study of cryotherapy for retinopathy of prematurity (ROP). Approximately one third of the eyes did not develop ROP, whereas two thirds developed mild-to-severe ROP. None of the eyes underwent cryotherapy. INTERVENTION: Refractive error was measured at 3 months, 1 year, and 5 1/2 years term due date at the five long-term follow-up centers. In most eyes, refractive error also was measured at 2, 3 1/2, and 4 1/2 years. MAIN OUTCOME MEASURE: Myopia was defined as 0.25 diopter (D) or greater with high myopia as 5 D or greater. RESULTS: The proportion of eyes with myopia in this preterm population was increased compared to published data on full-term children and was related to severity of both acute-phase and cicatricial-phase ROP. The percentage of eyes with myopia varied little across ages, ranging from 21.2% at 1 year to 15.7% at 4 1/2 years. The percentage of eyes with high myopia doubled from 1.8% to 3.9% between 3 months and 1 year and remained stable thereafter. The distribution of refractive errors in eyes with mild acute-phase ROP was similar to that of eyes with no ROP. In contrast, eyes with moderate or severe acute-phase ROP showed an increased prevalence of high myopia. The distribution of refractive errors changed between 3 months and 1 year with little change after 1 year. This pattern of refractive development differs from that of full-term infants. Birth weight, severity of ROP, and degree of myopia at 3 months predicted the presence of myopia and high myopia at 5 1/2 years of age. CONCLUSIONS: The distribution of refractive errors in preterm infants from age 3 months to 5 1/2 years varies with severity of acute-phase ROP and cicatricial disease. Changes in refractive error distribution occur primarily between 3 months and 1 year and involve a decrease in the proportion of eyes with hyperopia and an increase in the proportion with high degrees of myopia.


PURPOSE: To evaluate the effect of cryotherapy on refractive error status between ages 3 months and 10 years in children with birth weights of less than 1251 g in whom severe retinopathy of prematurity (ROP) developed in one or both eyes during the neonatal period. DESIGN: Randomized clinical trial. PARTICIPANTS: Two hundred ninety-one children in whom severe ROP developed during the neonatal period. INTERVENTION: Cryotherapy for ROP. MAIN OUTCOME MEASURES: Cycloplegic Refraction METHODS: The children underwent repeated follow-up eye examinations, including cycloplegic retinoscopy, between 3 months and 10 years after term due date. Refractive error data from all
eyes that were randomized to cryotherapy were compared with data from all eyes that were randomized to serve as controls. Refractive error data were also compared for a subset of children who had both a treated and a control eye that could be refracted. RESULTS: At all ages, the proportion of treated eyes that were unable to be refracted because of retinal detachment, media opacity, or pupillary miosis was approximately half the proportion of the control eyes that were unable to be refracted. When data from all eyes that could be refracted were considered, the distribution of refractive errors between fewer than 8 diopeters (D) of myopia and more than 8 D of hyperopia was similar for treated and control eyes at all ages. The proportion of eyes with 8 D or more of myopia was much higher in treated than in control eyes at all ages after 3 months. In the subset of children who had a treated eye and a control eye that could be refracted, distributions of refractive errors in treated versus control eyes were similar at most ages. CONCLUSIONS: In both treated and control eyes, there was an increase in the prevalence of high myopia between 3 and 12 months of age. Between 12 months and 10 years of age, there was little change in distribution of refractive error in treated or control eyes. The higher prevalence of myopia of 8 D or more in treated eyes, as compared with control eyes, may be the result of cryotherapy's preservation of retinal structure in eyes that, in the absence of cryotherapy, would have progressed to retinal detachment.


Quantitative electron microscopic analysis reveals 2.85 million retinal axons in fetal rhesus monkeys--a number that is more than twice the 1.2 million present in the adult. More than 1 million supernumerary optic axons are eliminated before birth, simultaneously with the segregation of inputs from the two eyes into separate layers of the lateral geniculate nucleus. Selective elimination of optic axons may not only play a role in the segregation of binocular visual connections but, secondarily, may establish the ratio of crossed and uncrossed retinogeniculate projections.


We report here that in mature rhesus monkeys in which one eye was removed during the first half of gestation, the optic nerve of the remaining eye is larger and contains significantly more retinal axons than in age-matched control animals. Such supernumerary fibres in monocularly enucleated monkeys probably result from an arrest in the normal process of elimination of excess embryonic optic axons. Although the function of retained supernumerary optic axons is unknown, this finding demonstrates that (1) competition between the two eyes for synaptic territory occurs prenatally, before visual experience and (2) an early lesion in one brain area can adjust or enhance the size and perhaps the performance of other synaptically related structures.


Monocular diplopia may be of external, optical, neurological, neuromuscular, or psychogenic origin. It may develop spontaneously or it may be induced by surgery or trauma. Because treatment is usually directed toward the cause, determination of the etiology is important. Useful diagnostic techniques are described, as are treatment modalities for monocular diplopia of various origins.


Objectives: To report the timing of involution of acute retinopathy of prematurity (ROP). Design: An analysis of prospective retinal observational data recorded at infants' eye examinations. Participants: Infants from the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity (CRYO-ROP) had birth weights less than 1251 g and served as subjects. The study population included 766 children who were examined in 5 of the 23 study centers and who developed at least 1 clock hour of acute ROP, stages 1 through 3. One eye from each patient was randomly chosen for analysis. Main Outcome Measures: Investigators documented the location, extent, and severity of ROP during serial retinal examinations. The onset of the ROP's involution was determined from a review of these data, applying a set of predetermined criteria. Results: Acute-phase ROP began to involute at a mean of 38.6 weeks postmenstrual age. In 90% of patients, the ROP began to involute before 44 weeks of postmenstrual age. Acute ROP that demonstrated involution by moving from zone II to zone III was associated with an unfavorable outcome in 2 (1%) of 200 cases. Retinopathy of prematurity that was present only in zone III during a child's serial retinal examinations was never associated with the development of a partial or total retinal detachment. Conclusions: The onset of involution of acute retinopathy of prematurity correlates better with postmenstrual rather than with chronological age. This is reminiscent of the reported similar correlation of postmenstrual age to the time of onset of prethreshold and threshold ROP. Zone III ROP was nearly always associated with a favorable outcome.

Visual defects are often poorly recognized in children with multiple neurologic problems due to perinatal hypoxic-ischemic encephalopathy. We report the clinical, radiologic, and electrodiagnostic characteristics of 20 children with cortical visual impairment secondary to birth asphyxia. Clinical diagnosis often was delayed. Ten patients recovered vision during the first two years of life. Four infants had coexisting damage to the pregeniculate visual pathway. Useful investigations included cranial computed tomography and visual evoked potential mapping. Electroencephalographic abnormalities were nonspecific. The classical definition of cortical blindness must be modified for children.

Retinoscopy was performed on a population of predominantly white esotropic children younger than 5.5 years with cyclopentolate 1% and atropine 1.0%. Atropine 1.0% revealed +0.34 diopters more hyperopia than cyclopentolate 1.0%, when the mean differences between the two drugs were examined. Mean difference analysis would probably indicate that atropine retinoscopy was unnecessary. However, 22% of the children had +1.0 diopters or more of hyperopia uncovered by atropine. This significant subpopulation suggests that in young patients with esotropia, an atropine refraction is essential to uncover the maximum amount of hyperopia. Almost all of this subgroup with +1.00 or greater hyperopia had an initial cyclopentolate retinoscopy of +2.00 diopters or more. Therefore, retinoscopy using atropine cycloplegia becomes even more important in this population. There was a trend for the greater differences to be in children older than age 2 years. However, these values were not statistically significant.

Fifty-six patients underwent adjustable rectus muscle recession procedure. This procedure permits the surgeon to enhance or diminish the amount of muscle recession on the evening after surgery or the first postoperative day if cover-testing indicates an inappropriate amount of undercorrection or overcorrection. The adjustable rectus muscle recession technique seems to be a practical and effective means to change the strabismic deviation postoperatively. The procedure requires patient cooperation and is most suitable for patients age 15 years and older. The procedure has been effective in altering the angle of deviation, and this alteration has been stable during the follow-up period in most cases. In this initial series, the reoperation rate was low, postoperative alignment was excellent, and complications were minimal.

Can the pinhold principle be practically applied to solving the problem of providing useful vision for aphakics without resorting to aphakic spectacles? The authors have experimentally quantified the pinhold effect and concluded that the pinhold, successfully produced by miotics in certain patients or by a thin, dyed contact lens, may have clinical value.


Chlamydia trachomatis was isolated from the most inflamed eye of 13 of 107 (12%) infants with neonatal purulent conjunctivitis and from none of 100 healthy infants (p less than 0.01). Staphylococcus aureus was recovered from 49 (46%) inflamed eyes and from 8 (8%) healthy eyes (p less than 0.01). Streptococcus pneumoniae, Haemophilus influenzae, Branhamella catarrhalis, Escherichia coli and Klebsiella pneumoniae were isolated from very few infants with conjunctivitis but not from controls. No organisms could be recovered from 23 (22%) infants with conjunctivitis and from 60 (60%) healthy infants (p less than 0.01). The incidence of neonatal purulent conjunctivitis was 107 (2%), of 5,924 births. Eyes infected with C. trachomatis were significantly more inflamed than eyes from which S. aureus or no organisms could be isolated. Furthermore, conjunctival "pseudomembranes" were associated with C. trachomatis. The age at onset of the chlamydial conjunctivitis was higher compared to the age at onset of conjunctivitis in which S. aureus or no organisms were isolated.

BACKGROUND: There exists no reliable information that allows the ophthalmologist to predict with any degree of certainty a particular infant's chances of requiring surgical treatment for retinopathy of prematurity (ROP) or of reaching an unfavorable outcome on the basis of the retinal findings at the time of the nursery examination. METHODS: In the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity (CRYO-ROP), 4099 infants weighing less than 1251 g at birth underwent eye examinations that began at 4 to 6 weeks after birth and subsequently continued at 2-week intervals. Independent variables in the population were studied using multiple logistic regressions. RESULTS: An increased risk of reaching threshold ROP was found associated with lower birth weights, younger gestational age, white race, multiple birth, and being born outside a study center nursery. For infants who developed ROP (66%), corresponding probabilities are presented for developing severe ("threshold") ROP or of having an unfavorable macular outcome. The risk of an unfavorable macular outcome was increased with zone 1 ROP, "plus" disease, the severity of the stage, and the amount of circumferential involvement. A higher risk also was associated with a rapid rate of progression of ROP to prethreshold disease but not with the postconceptional age at which ROP was first noted. CONCLUSION: The findings indicate that the
oculomotor characteristics of ROP, along with some easily identifiable and available basic systemic and demographic information about an infant, can assist the ophthalmologist in understanding variations in an individual baby's chance for a good or poor macular outcome.


Visual acuity, visual fields and neurological status were assessed in 10 infants with periventricular leukomalacia (PVL), tested at 16, 36, 48 and 72 weeks from the expected date of confinement. Monocular acuity development was normal in eight of the 10 infants, but was below normal in one infant at eight months and in another at 18 months. Over half the infants tested at 16, 36 and 48 weeks had smaller visual fields than those of 95 per cent of healthy preterm infants tested at the same ages, but by 72 weeks only two of six infants tested had restricted visual fields. Nine of the 10 infants were neurologically abnormal at ages under one year, but only four remained so beyond one year. These results indicate more favourable outcomes for visual acuity and neurological status in infants with non-cavitary PVL than have been reported in infants with cavitary PVL. The most compromised infants were one with cavitary PVL and another with extensive non-cavitary PVL who had the longest-lasting EEG abnormalities of all 10 infants.


Sixty-seven injections of botulinum. A toxin were given to patients for correction of strabismus. No systemic complications of any kind have occurred. The maximum time of paralysis occurs four to five days following the injection, and then gradually diminishes, depending on the dose. The maximum correction of strabismus has been 40 prism diopters. The maximum follow-up following injection is six months. Injection of botulinum A toxin into extraocular muscle to weaken the muscle appears to be a practical adjunct or alternative to surgical correction.


BACKGROUND: Radioactive episcleral plaque brachytherapy is a treatment method for selected retinoblastomas. The authors have used this technique since 1976 as both a primary and a secondary treatment method after other methods failed to achieve tumor control. METHODS: A review of the records of 400 consecutive children with retinoblastoma showed that solitary plaque radiotherapy was used as a method of management in 103 cases. The authors' overall experience was evaluated, and the results between primary and secondary plaque therapies were compared in these 103 cases. RESULTS: Of the 103 tumors, the mean basal diameter was 7 mm, and the mean thickness was 4 mm. Overlying vitreous seeds were clinically apparent in 50 cases (48%). The mean proximity of the tumor margin to the optic disc margin was 6 mm and to the foveola was 6 mm. The mean follow-up period was 40 months. In 89 cases (86%), the tumor was controlled by one plaque application, whereas in 13 cases (13%), tumor recurrence after initial tumor shrinkage necessitated subsequent treatment. Final visual outcome was good in 63 cases (62%), poor in 30 (29%), enucleation in 9 (9%), and unknown in 1 case. The poor vision was due to foveal retinoblastoma (with or without amblyopia) in 25 cases (83%). Eight of the nine enucleated eyes were treated initially with external beam radiotherapy then later with plaque radiotherapy. In 31 cases (30%), plaque radiotherapy was used as a primary treatment to the tumor, while in 72 cases (70%), it was a secondary form of management after failure of other methods to control the tumor. Statistical analysis showed that tumors treated with plaque radiotherapy as a primary measure were more likely to be larger in in base (P = 0.01) and thickness (P = 0.01) than secondary treated tumors. The secondary treated retinoblastomas were more likely to have vitreous seeds (P = 0.02) than the primary treated tumors. The rate of tumor control and patient survival was similar between the two groups. CONCLUSION: Plaque radiotherapy is very effective in treating selected retinoblastomas with a high rate of tumor control and patient survival. It can be used successfully as a primary or a secondary treatment for tumors that have not been adequately controlled by other therapeutic methods.


A review of 400 consecutive patients with retinoblastoma disclosed that 103 tumors in 103 eyes were treated with solitary plaque radiotherapy. The tumors were from 1 to 16 mm (mean, 7 mm) in basal diameter and from 1 to 8 mm (mean, 4 mm) in thickness. Thirty-one tumors were treated with plaque radiotherapy as initial treatment, whereas 72 tumors were treated with plaque radiotherapy as secondary treatment after failure of other methods. Of the 102 tumors on which adequate follow-up data were available, all responded initially to plaque radiotherapy with tumor regression. Over the mean follow-up of 38 months (range, six to 192 months), 89 tumors (87%) showed persistent regression and 13 (13%) showed tumor recurrence. The recurrence occurred at a mean interval of five months (range, one to 11 months) after plaque radiotherapy. A statistical analysis of tumor size, tumor location, tumor proximity to the optic disk and foveola, presence of vitreous seeds, radioactive plaque diameter, plaque shape, radioisotope, and primary or secondary treatment disclosed no important predictors of tumor recurrence. Carefully selected retinoblastoma, even
juxtapapillary and macular tumors and those with localized vitreous seeds, can be successfully treated with plaque radiotherapy.


For many years, enucleation has been the most common treatment of retinoblastoma. It generally has been done on the affected eye of children with unilateral disease and on the more severely affected eye in children with bilateral disease. In recent years, there has been a trend toward earlier diagnosis of retinoblastoma, and there have been refinements in conservative therapeutic methods, such as radiotherapy, photocoagulation, and cryotherapy. Consequently, it often is possible to salvage the affected eye and retain useful vision in many patients who would have undergone enucleation in the past. This article provides an update on the diagnosis and treatment of retinoblastoma, with emphasis on certain misconceptions related to the management of this tumor. New genetic aspects and their relationship to counseling are discussed briefly.


A review was done of 250 consecutive biopsies for orbital space-occupying lesions in children. Benign cystic lesions were by far the most common, accounting for 52% of the lesions. Inflammatory lesions, usually biopsied to exclude the diagnosis of rhabdomyosarcoma, accounted for 16%. Among the 250 biopsies there were 14 primary malignant tumors (6%), ten of which were rhabdomyosarcoma. Although rhabdomyosarcoma is not the most common space-occupying orbital lesion in children, clinicians should be familiar with its clinical features and proceed with immediate biopsy followed by appropriate irradiation and chemotherapy if the diagnosis is established. However, biopsy should only be undertaken if there is reasonable suspicion that the lesion could be rhabdomyosarcoma or other childhood orbital malignancy.


Of 500 consecutive patients referred to the Ocular Oncology Service at Wills Eye Hospital with the diagnosis of possible retinoblastoma, 288 (58%) were found on clinical evaluation to have retinoblastoma and 212 (42%) had lesions that simulated retinoblastoma. A total of 23 different conditions accounted for the 212 pseudoretinoblastomas. Three most common pseudoretinoblastomas were: persistent hyperplastic primary vitreous (28%), Coats' disease (16%), and presumed ocular toxocariasis (16%). Congenital cataract and retinopathy of prematurity accounted for a much lower percent of pseudoretinoblastomas in this series as compared to a previously reported series. The pertinent clinical features that serve to differentiate these simulating lesions from retinoblastoma are reviewed.


It is important for the pediatrician to be aware of the benign and malignant ophthalmic tumors that can occur in children. This article covers some general concepts related to ocular tumors in children and briefly describes the clinical features and management of some of the more important tumors of the eyelids, conjunctiva, intraocular structures, and orbit. Although most ocular tumors in children are benign, certain malignant neoplasms can threaten the child's life as well as the child's sight. Hence, prompt recognition and patient referral to an ocular oncologist are crucial for proper clinical management.


For many years the most common treatment for retinoblastoma has been enucleation, generally performed on the affected eye in children with unilateral sporadic disease and on the more severely affected eye in children with bilateral disease. With refinements in conservative treatment methods, however, the affected eye now often may be salvaged and useful vision retained. Emphasizing this trend, we present our current approaches to managing retinoblastoma based on our experience with 324 patients, outlining our indications and pointing out a number of misconceptions about the role of enucleation, photocoagulation, cryotherapy, and radiotherapy in treating this condition. We also sketch some recent findings regarding the genetics of retinoblastoma and consider ways in which such research may lead to improved management of the disease.


Retinoblastoma is a malignant intraocular tumor of childhood that requires accurate diagnosis and prompt treatment. It is well known that several other ocular conditions of childhood can clinically simulate retinoblastoma. Knowledge of the clinical features that serve to differentiate retinoblastoma from simulating lesions may assist the clinician in arriving at the correct diagnosis and preventing misdirected therapy. This review provides a simple classification of those pseudoretinoblastomas and reviews the recent literature on this subject. It emphasizes the clinical features that help differentiate pseudoretinoblastomas from true retinoblastoma.


Three dimensional reconstruction, with the use of serial, 1-micrometer sections, has revealed a system of oriented intercellular spaces within the undifferentiated optic cup. These large openings appear in the marginal zone of the primitive retina and optic stalk prior to the formation of the first retinal ganglion cell axons. The spaces at the region of the optic disc form sets of long, interconnecting tunnels oriented in the direction of the stalk. The spaces at the back and
rim of the cup form blind, radially arranged pockets. The extracellular tunnels of the optic disc region strictly maintain their positions in relation to the optic fissure and, thus, discrete portions of the retina become connected by continuous openings with equivalent regions in the stalk. The path taken by the earliest outgrowing optic fibers is identical to the one previously established by the intercellular tunnels. We propose that the tunnel and pocket layout may provide directional and topographic information to the first forming optic axons.


Optic nerve hypoplasia is frequently associated with congenital CNS anomalies and endocrine disturbances. In a prospective study of 93 children with this condition, we found that 78% of those with bilateral involvement, poor vision, and nystagmus (group 1) had additional nonocular developmental abnormalities. In contrast, among children with unilateral optic nerve hypoplasia (group 2) or with bilateral segmental hypoplasia and good vision (group 3), only 21% had nonocular developmental abnormalities. Delayed development in 23 children was the most frequent nonvisual problem associated with optic nerve hypoplasia, and in five cases it occurred in the absence of detectable CNS, endocrine, or medical abnormalities. Nine patients had decreased levels of thyroid hormone; three, decreased levels of growth hormone; and three, decreased levels of both. Five had histories of neonatal hypoglycemia. Of the children with endocrine disturbances, ten had delayed development, but only seven had anomalies on computed tomographic scan.


Thirty-four patients (49 eyes) who had dissociated vertical deviation were treated with faden operation of Cuppers. The median follow-up period was 14 months. The operation produced good results in 23 of the patients, some improvement in seven, and did not help four. The posterior fixation suture was placed 14 mm behind the superior rectus muscle insertion and the superior rectus muscle was recessed.


Retrospectively studied were 91 patients, 14 years of age or less, who had 152 penetrating keratoplasties in 107 eyes, with an average follow-up of 30.1 months. Survival analysis showed the probability of obtaining a clear graft at one year to be 60 +/- 8% in 45 eyes with congenital opacities, 70 +/- 8% in 31 eyes with opacities from trauma and 73 +/- 8% in 31 eyes with acquired nontraumatic opacities. Most failures occurred during the first postoperative year and were characterized by the gradual loss of graft clarity from undetermined cause. Twenty-nine percent (10) of the 34 eyes with congenital corneal opacities in which vision could be measured had a most recent visual acuity better than 6/120 (20/400). Irreversible amblyopia, glaucoma, other structural abnormalities of the anterior segment and mental retardation complicated visual rehabilitation in the congenital group. In the post-traumatic group, vision was better than 6/120 (20/400) in 45% (13) of 29 eyes, being limited by vitreoretinal pathology, fibrous ingrowth, and optic nerve damage from glaucoma. Visual acuity in the acquired, nontraumatic group was better than 6/120 (20/400) in 67% (20) of 30 eyes. The percentage of patients achieving 6/12 (20/40) or better in the congenital, post-traumatic, and nontraumatic groups were 3% (1/34), 17% (5/29), and 47% (14/30), respectively. In the congenital group, only patients with posterior polymorphous dystrophy obtained a visual acuity of 6/30 (20/100) or better. Preoperative vascularization of the cornea, persistent epithelial defects, and performance of lensectomy-vitrectomy were factors most highly correlated with poor graft survival.(ABSTRACT TRUNCATED AT 250 WORDS)


A 14-month-old boy with developmental delay showed microcephaly, spastic diplegia, central visual fixation and an esotropia. A head tomographic scan disclosed absence of normal occipital cortex and electroencephalography showed markedly reduced voltages over the occipital region. Visual development in this patient may be related to heterotopic occipital cortex of a functioning non-striate system of visual processing. Visual function cannot be predicted when severe developmental anomalies of the occipital cortex are detected with computerized tomography.


Few data exist from which an optimal strategy for retinopathy of prematurity (ROP) screening can be deduced. This strategy should compromise between the dual purpose of screening, to wit monitoring the incidence of ROP and determining an optimal moment for therapeutic intervention. We recorded the timely incidence and course of ROP in all except four premature infants admitted during a 1-year period. The study results indicate that a single screening moment is not likely to detect more than 70% of all cases. When relying, however, on a single screening, the investigation should be performed between the 7th and 9th weeks. The unpredictable course of ROP, varying between early starting, rapidly progressing forms and very late starting, usually benign forms probably precludes the possibility of 100% ROP detection. Screening during the 4th to 5th, 8th, and 11th weeks probably gives the best chances for determination of an optimal moment for treatment and approximation of the incidence.


Bruckner, in 1962, published a paper in German describing a "trans-illumination" test extremely useful in the diagnosis of small angle deviations and amblyopia in young uncooperative children. A bright coaxial light source, such as a direct ophthalmoscope, is used. Both eyes of the patient are simultaneously illuminated from approximately one meter distance. First, the position of the corneal light reflex, (Hirschberg test), along with brightness difference of the fundus reflex as seen in the pupil through the ophthalmoscope, is evaluated. When strabismus is present, the fixing eye has a darker reflex than the deviated eye. The second step evaluates pupil size, pupil reaction, and fixation movement of the eyes on "successive" illumination of one eye at a time. This is useful in detecting amblyopia.


Retroequatorial fixation of a rectus muscle by means of nonabsorbable scleral sutures weakens this muscle in its primary field of action without disturbing the balance with its antagonist in other positions of gaze. This procedure has been employed in patients with dissociated vertical deviations, nystagmus compensation (blockage) syndrome, double-elevator paresis, nonaccommodative convergence excess, and Duane's retraction syndrome. Complications were not encountered, but the operation was ineffective in some patients and reoperation became necessary.


Repertoire gonioscopy of children with congenital aniridia confirms the presence of an angle abnormality which can be progressive and cause glaucoma. This abnormality features obstruction of the trabecular meshwork by variable mixtures of anterior migration of the peripheral iris and thickening of the uveal meshwork associated with a vascular net over exposed trabecular meshwork adjacent to the anterior edge of the iris. Preliminary results of prophylactic gonio-surgery in 28 eyes of 16 children with an average age of 4 years was reported. This surgery was performed without complication and produced a permanent exposure of the trabecular meshwork to the anterior chamber for an average of 8 circumferential hours, if two procedures were performed. Preliminary results suggest a stabilization of eye pressures at least through childhood and encourage the continuation of these prophylactic operations on selected eyes with congenital aniridia. Therapeutic goniotomy for established acquired glaucoma in congenital aniridia cannot be relied on, but may be a benefit for early detected cases or for glaucoma associated with aniridia in infancy.


This paper classifies the abnormalities of the anterior chamber cleavage syndrome (mesodermal dysgenesis of the iris and cornea). The anatomic findings are arranged in a tabular stepladder fashion which builds from simple to more complex combinations, most of which have been previously known by eponyms. There are three groups of anomalies: 1) peripheral, 2) central, and 3) combinations of the two. 1) The peripheral anomalies consist of a prominent Schwalbe's
ring, iris strands to Schwalbe's ring, and hypoplasia of the anterior iris stroma. Developmental glaucoma is commonly present. 2) The essential feature of the central anomalies is a defect in the corneal endothelium and Descemet's membrane with an overlying corneal opacity. Additional components include central iridocorneal adhesions, keratolenticular approximation with cataract, and scleralization of the cornea. Chorioretinal anomalies, developmental glaucoma, and systemic malformations may be present. 3) Central and peripheral combinations may exist in the same eye, in both eyes of the same patient, or within the same family.


Whiting, S., et al. (1985). "Permanent cortical visual impairment in children." Dev Med Child Neurol 27(6): 730-739. Fifty patients with permanent cortical visual impairment were evaluated. They had a characteristic behaviour profile, usually with residual sight but poor visual attention. 30 of the 50 also had damage to the anterior visual pathway. Visual evoked potential mapping was shown to have a clear advantage over visual evoked responses, and using that in conjunction with CT and clinical data enabled several subgroups of cortical visual impairment to be identified. The diagnosis probably is more common than previously recognised, and should be suspected when there is greater delay in visual development in other areas and the degree of visual loss is unexplained by ocular findings. Using traditional criteria for cortical blindness may mean that many children are not diagnosed, which has serious implications for their rehabilitation.


Although prophylactic eyedrops to treat ophthalmia neonatorum is mandated nationwide, states and hospitals are free to choose specific drugs. To compare two of these agents, we studied the incidence and characteristics of ophthalmia neonatorum in two UCLA teaching hospitals over a five-year period. One, which used 1% silver nitrate solution exclusively, had 50 cases in 34,772 births, a frequency of 0.14%. The other used 0.5% erythromycin ointment exclusively and had 43 cases in 12,652 births, a frequency of 0.34%. Ophthalmia neonatorum was more frequent in the hospital using erythromycin (p less than 0.001), as was chlamydial conjunctivitis (p less than 0.02). Although not statistically significant, gonococcal conjunctivitis was found in four infants, all in the hospital using silver nitrate. Because silver nitrate was found more effective in decreasing the total frequency of all cases of ophthalmia neonatorum, and cases caused by Chlamydia and gram-negative bacteria specifically, this drug still should be considered as a primary prophylactic agent against ophthalmia neonatorum.


The anatomic and visual results of vitreous surgery in 140 eyes of 108 children with stage 4 and 5 retinopathy of prematurity (ROP) were reviewed. Macular attachment was achieved in 9 (64%) of 14 eyes with stage 4. In stage 5 eyes, partial posterior attachment was obtained in 38 (31%) of 121 eyes and complete posterior attachment was obtained in 11 eyes (9%). Final visual acuity of fix and follow or greater occurred in 6 (43%) of 14 eyes with stage 4 and in 13 eyes (11%) with stage 5 ROP. Retinal detachment (RD) funnel configurations were classified according to the International Committee on ROP. The funnel type which was wide anteriorly and wide posteriorly had the best anatomic (63%) and visual (19%) success. Reoperation rarely improved visual acuity (4%), although reattachment was possible in 6 (22%) of 27 eyes. All children in this series underwent surgery by 2 years of age. Anatomic and visual success was achieved less often in children who underwent surgery at age 6 months or younger.