

STAGE 5 RETINOPATHY OF PREMATURETY IN ONE EYE – CASE REPORT*Gordana Stanković-Babić¹, Ana Oros², Milena Vujanović³ and Sonja Cekić³*

Retinopathy of prematurity (ROP) is a leading cause of blindness in children and one of the most important reasons of blindness in the perinatal period.

The aim of the paper was to present a nine-month-old baby boy with esotropia, microphthalmos and completely detached retina in one eye, as the end stage of the disease, who had not been checked for ROP. The boy was born in the 32nd gestational week, with 1670 g birth weight. Indirect ophthalmoscope examination and ultrasonography of the left eye showed stage 5 retinopathy of prematurity. On the right eye, the finding was valid.

Retinopathy of prematurity today needs recognition, understanding and awareness among ophthalmologists, pediatricians, neonatologists. Early diagnosis of damage is important in the treatment of ROP. *Acta Medica Medianae* 2012; 51(1):37-41.

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Introduction

Retinopathy of prematurity (ROP) has been increasingly recognized as an important cause of childhood blindness in industrialized and developing countries (1). It is a very serious disorder, previously known as retrolental fibroplasias (Terry, 1942) (2), which is characterized by abnormal vascular development of retina in premature infants (3). Low gestational age (especially <32 weeks of gestation), low birth weight (<1500g, especially <1250g), sepsis and oxygen therapy are dominant risk factors for this condition (4-11). Neonatal sepsis, oxygen exposure and low gestational age may have synergistic effects, while oxygen exposure and sepsis may have antagonistic effects on developing ROP (11). Also, high blood concentrations of oxygen and carbon dioxide (9,12) and low blood pH are associated with increased risk of severe ROP (12). Other risk factors for ROP are concurrent illness (9), heart disease, infection, apnea, respiratory distress, bradycardia, white race, intraventricular hemorrhage, prolonged parenteral nutrition, lung maturation, steroid treatment, blood transfusions (4,10) and maternal factors such as preeclampsia, heavy

smoker, older maternal age, multiple births, in vitro fertilization (10) etc.

The International Classification of Retinopathy of Prematurity (ICROP) uses a number of parameters to describe the disease. Diseases are classified according to zones they affect (1, 2, and 3), the circumferential extent of the disease based on the clock hours (1-12), the severity of the disease (stage 1-5) and the presence or absence of "Plus Disease" (13). Stage 5 is the severest form of ROP, the final stage of the disease and the main cause of visual impairment and blindness. This stage is a total retinal detachment in the shape of a funnel - stage 5A is an open funnel and stage 5B is a closed funnel. However, even milder forms of ROP leave ocular sequel in the form of ametropias, refractive errors (especially myopia), strabismus, glaucoma, cataracts (14).

Treatment for ROP might include: a) cryotherapy (freezing) to prevent the spread of abnormal blood vessels; it was the original mode of treatment since the 1970s.; b) Laser surgery (eg, xenon, argon and diode) has shown to be as effective as cryotherapy for ROP; c) Scleral buckling surgery and/or vitrectomy was performed for stages 4 and 5, but with the poor anatomical and visual prognosis (4, 15,16).

In January 2008, the implementation of the screening of premature babies for ROP in the region of Niš started. All premature neonates included in the examination had birth weight of 2000g or less and/or gestational age of 37 weeks or less; also, there were other neonates born after the 37th gestational week, however, with some other associated risk factors (oxygen, ventilation, sepsis, etc).

The aim of our study was to present a child with ROP stage 5 and microphthalmus on the same eye, which had not been previously checked for ROP.

A case report

A case of a nine-month-old preterm child, of male sex presented with deformed head due to the previous operations for hydrocephalus and with an asymmetrical face. The left eye globe was smaller than the right one, with an esodeviation. The parents brought the child to the Children's Pediatric Clinic because of the left eye sliding. The child was referred from the center where screening for ROP had not been performed.

The baby is the first child from the first regular pregnancy. The mother (22 years old) was a heavy smoker. A preterm baby, 32 weeks of gestation, birth weight 1670g, with a respiratory distress syndrome, icterus, APS 6/7, oxygen therapy. The subsequent controls found hydrocephalus for which the child had been operated on two occasions when the pumps were installed. Family history was negative. A regular finding was obtained after performing a dilated retinal examination.



Figure 1 Microphthalmus and esotropia

Ophthalmic examination of anterior segment showed a small left eye globe, with an esodeviation, moving in all directions, but did not follow the light (Figure 1.). The red reflex was present only on the right eye. Leukocoria was present on the left eye (Figure 2). Corneal diameters were 9.5 mm on the right eye and 8 mm on the left eye. Electro diagnostic tests were undertaken - VEP on the right eye showed weaker cortical responses, amplitude and latency in physiological limits. Neovascularization of the iris on both eyes was absent. The child was examined by indirect ophthalmoscope with +20 D lens with a full dilatation of the pupils (sol.Tropicamid 0.5% and sol. Phenylephrine 1% combination drops). Examination of anterior and posterior segments of the right eye was normal. Computerized refractometry (Righton Speedy-K, Japan) showed the presence of oblique astigmatism on the right eye +0.75/+1.75 160°. The measurement of axial length by ultrasonography A scan were Lax = 17.5 mm right

eye (Figure 3). B scan - total ablation of the retina on the left eye which was smaller (Figure 4). Electrodiagnostic tests, such as VEP, were undertaken. The right eye showed weaker cortical responses, amplitude and latency in physiological limits. Genetic examination showed normal male karyotype.

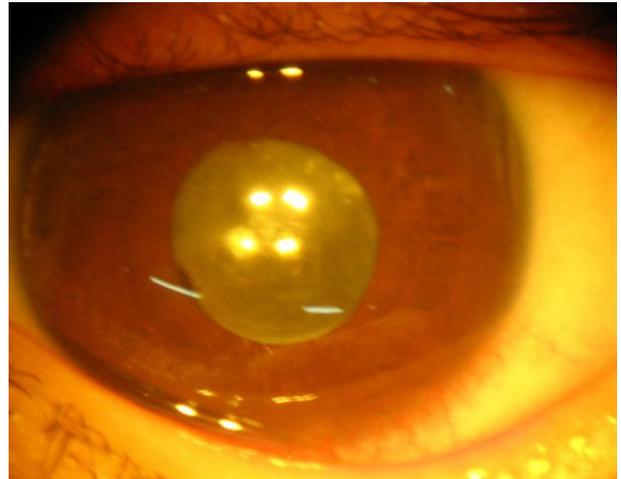


Figure 2 Leukocoria of the left eye

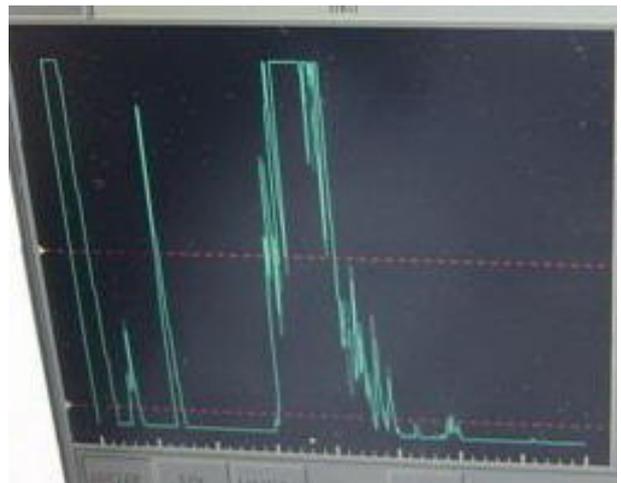


Figure 3 A scan of the right eye

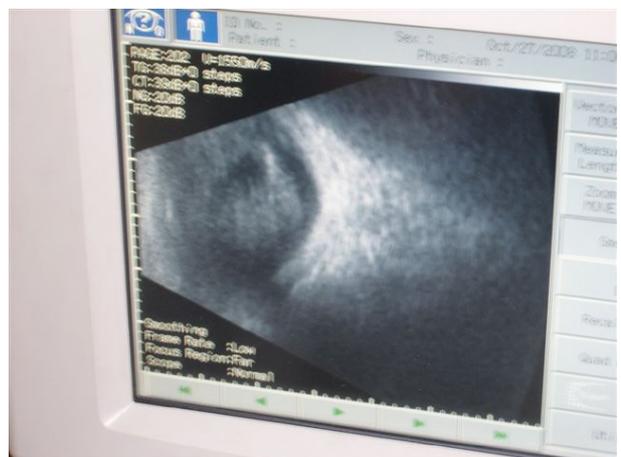


Figure 4 B scan of the left eye

The therapy involved the correction of the right eye +0.25 /+ 0.75 160°, and eye prosthesis (the cosmetic aspect and the social interaction of the patient) in future for the left eye.

The neurologist was satisfied with the findings at the last follow-up examination. The parents were introduced to the nature of the disease.

Discussion

Retinopathy of prematurity is important (1,2), but a potentially preventable cause of childhood blindness (17).

The introduction of intensive care units into neonatal nurseries has caused a significant increase in the survival rate of infants of very low birth weight (500-1500g) with significant increase of ROP (18). In general, more than 50% of premature infants weighing less than 1250 g at birth show evidence of ROP, and about 10% of the infants develop stage 3 ROP. The incidence of ROP is more frequent in white infants than in black infants and in male infants than in female infants (19).

The unique feature of ROP relates to its occurrence only in premature infants with immature and incompletely vascularised retina (16). The development of the human retinal vasculature commences at approximately the 16th week of gestation and concludes at term (i.e., the 40th week of gestation) (20). Understanding both the mechanisms of normal retinal vascular development and the pathophysiological processes ROP is the key to developing new therapeutic approaches to prevent complications of the ROP (21).

A promising future strategy to counter ROP is to use anti-VEGF therapy (VEGF-specific neutralizing antibody, bevacizumab). Timing (stage/zone of ROP), dose (0.4–12.5 mg intravitreal), and frequency of administration of bevacizumab, as well as a co-treatment with photocoagulation, varied tremendously among reports (22). In the future, evaluation of candidate genetic polymorphism influencing the outcome of ROP may provide new information about the pathogenesis of the disease. Screening of genetic polymorphisms may also help to identify and treat those infants who are at high risk in a timelier manner (19).

Early diagnosis of damage is important in the treatment of ROP. In November 2009, on a World ROP Congress in New Delhi, screening criteria for premature babies were modified. The first ophthalmology exam depending on the baby's gestational age should be 1-3 weeks after birth. Babies born at 34 weeks or later have their exam at 1 week after birth. Those born earlier have exams later at 2 weeks (31, 32 and 33 gestational age) or 3 weeks after birth, for a baby of a very low gestational age. Follow-up examinations are determined based on the results of the first exam. Babies do not need

another examination if the blood vessels in both retinas have completed normal development.

Early treatment has shown to improve a baby's chances for normal vision. Laser treatment should start within 72 hours of the eye exam. Some babies with "plus disease" need immediate treatment (15,16).

Approximately 20% of all premature babies can develop some form of strabismus or refractive error by the time. This is why babies who are younger than 32 weeks or less than 1500 g receive follow-up care every six months, whether or not ROP is present (19). During the first months of life, preterm infants demonstrated more myopia, astigmatism and anisometropia than full-term infants did (13,18,23-28). Recommendations for follow-up examinations must include all aspects of visual function, i.e., visual acuity, contrast sensitivity, visual fields, refraction, strabismus, and perceptual problems (24).

Children born prematurely were reported to have an increased incidence of visual impairment, because of retinopathy of prematurity, but also because of perinatal lesions in the brain (29).

Microphthalmus is a congenital ocular anomaly. There is no predilection with respect to gender or race; it can be simple or complex (30), unilateral or bilateral, in over 50% of cases it is associated with systemic abnormality. Epidemiological data indicate the following risk factors: maternal age (over 40 years of age), multiple pregnancies, children low birth weight, low gestational age (31), prenatal infection of rubella, cytomegalovirus, fetal alcohol syndrome (32) and others.

We present a child with ROP stage 5 and microphthalmus on the same eye, who had not been previously checked for ROP. Risk factors for ROP are: prematurely born infant, low gestational age, a respiratory distress syndrome, icterus, oxygen exposure, hydrocephalus for which the child was operated on two occasions, young age of a mother who was a heavy smoker. Risk factors for microphthalmus are low birth weight and low gestational age of a child. The differential diagnostic problems that must be observed in a presented case involve persistent hyperplastic primary vitreous (PHPV) or persistent fetal vasculature (PFV) as a developmental ocular abnormalities consisting of a varied degree of glial and vascular proliferation in vitreous cavity. It is usually associated with a slightly small eye. Retinal detachments may be seen, rarely bilateral. Familiar exudative vitreoretinopathy (FEVR) may present with peripheral retinal capillary without perfusion; it may be bilateral and asymmetric, with strabismus or leukocoria in childhood. Phthisis is possible (9). Though being similar to ROP in our patient, there should be positive family history and there should be no history of prematurity or oxygen therapy.

The paper highlights the importance of timely screening for ROP in the prevention of blindness due to ROP stage 5, which fortunately developed only one eye in our small patient, in combination with microphthalmus.

Effective strategy for decreasing ROP-related blindness is performing retinal examinations in neonatal intensive care units. All interested parties must cooperate in developing and implementing full proof screening protocols. Hospital officials, nursery personnel, neonatologists and ophthalmologists all have areas of responsibility in ensuring adequate screening. Future safe and nondestructive therapeutic strategies combined with preventive approaches

need to be tailored to the unique developmental requirements of the premature infant (23).

Conclusion

Retinopathy of prematurity is the major ocular disorder of the neonate and the dominant cause of severe visual impairment in childhood. Prevalence of ROP and blindness will be significantly higher in infants with late retinal examinations. Prevention includes adequate prenatal care which minimizes premature birth and appropriate systemic intensive care which lessens the tissue hyperoxia /hypoxia swings. Early diagnosis of damage is important in the treatment of ROP.

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PETI STADIJUM RETINOPATIJE PREMATURITETA NA JEDNOM OKU - PRIKAZ BOLESNIKA

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Prematurna retinopatija (ROP) je vodeći uzrok slepila kod dece i jedan od najznačajnijih razloga slepila u perinatalnom periodu.

U radu se prikazuje dečak uzrasta devet meseci sa ezotropijom, mikroftalmusom i totalnom ablacijom mrežnjače na jednom oku kao poslednjim stadijumom bolesti, a koji nije prošao rutinski pregled na ROP. Dečak je rođen u 32. gestacijskoj nedelji, telesna masa na rođenju bila je 1670 g. Indirekna oftalmoskopija i ultrasonografija levog oka pokazale su peti stadijum prematurne retinopatije. Na desnom oku nalaz je bio normalan.

Prematurna retinopatija danas zahteva prepoznavanje, razumevanje i saradnju između oftalmologa, pedijatara, neonatologa. Rana dijagnoza oštećenja je značajna u tretmanu ROP-a. *Acta Medica Medianae* 2012;51(1):37-41.

Ključne reči: retinopatija prematuriteta, ablacija mrežnjače, slepilo, mikroftalmus