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CASE REPORT

Retinopathy of prematurity – developing too soon in babies born too early : A report of three cases

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Abstract: Nigeria has the third largest incidence of premature birth, after India and China. Most babies that will require treatment for retinopathy of prematurity (ROP) have birth weight (BW) less than 1500g and gestational age (GA) less than 32 weeks, with initial eye examination within the first 30 days of life. This report aims to present the findings in three babies with vision threatening ROP that developed at less than 30 days of life.
Routine screening for ROP was conducted for three babies. One was born GA 33 weeks, BW 1.6kg, screened for ROP on the 13th day of life, while the other two were a set of twins born at GA of 33 weeks, both BW 1.3kg, screened for ROP first on the 16th day of life. The twins were found to have arborizing vessels at the initial exam, which progressed to stage 3 pre-plus ROP by the 23rd day of life. The 3rd child had stage -3 plus disease at the first screening, warranting immediate LASER therapy.
The screening criteria of Nigeria ROP group is BW ≤ 1500g or GA of ≤ 34 weeks, with the first eye examination within the first 30 days of life. These case reports show that larger babies can also develop sight threatening ROP within the first two weeks of life. There is a need for constant review of ROP screening criteria and timing based on locally available data.

Keywords: Retinopathy of prematurity, screening criteria, AP-ROP

Introduction

Nigeria has the third largest incidence of premature birth, after India and China.[1]Over the last decade ROP screening in Sub Sahara Africa, particularly in Nigeria, has improved following several national and international collaborative efforts.[2]Most babies that will require treatment for ROP have birth weight (BW) less than 1500g and gestational age (GA) less than 32 weeks. Initial eye examination is usually done within the first 30 days of life because ROP is not considered present at birth. The screening criteria of Nigeria ROP group is BW ≤ 1500g or GA of ≤ 34 weeks,[2]with the first eye examination within the first 30 days of life. This report aims to present the clinical findings of three babies with vision threatening ROP that developed at less than 30 days of life.

In our hospital, routine ROP Screening is carried out just before discharge of babies from the neonatal intensive care unit (NICU) or at four weeks postnatal age, whichever comes first. We have found screening before discharge to be helpful in reducing cases of babies who miss screening, and so that has been established as part of our local guidelines. The screening for ROP is conducted using the Phoenix GO ICON retinal camera (Phoenix Clinical, Inc., Pleasanton, CA, USA). There were three babies born at 33 weeks gestational ages, referred to our tertiary facility having been born at peripheral hospitals, one being a single birth while the other two were twins. The NICU is a Level II nursery with facilities for mechanical ventilation and monitoring of oxygen saturation with pulse oximeters but no oxygen blenders. Oxygen is delivered in our NICU sometimes as piped oxygen, at other times from the concentrator depending on availability, at a flow rate of 2-3L/min with oxygen saturation ranging from 80-95%. Oxygen saturation is usually measured six hourly by the nurses. All the children in this series received phototherapy for physiologic jaundice. These babies were screened for ROP earlier than 30 days of life because they were going to be discharged from the NICU.

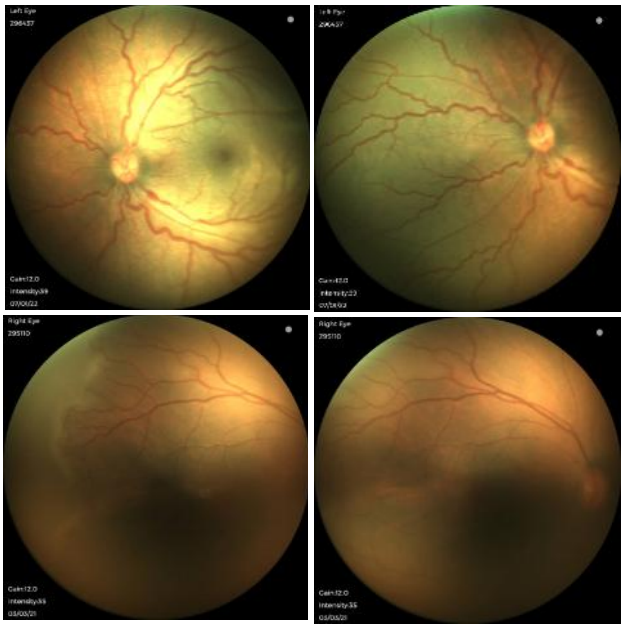
Case Report

Case 1

The first baby was admitted at the postnatal age of 48 hours on account of respiratory distress. Weight on ad-

mission was 1600g. The child had dysmorphic features consistent with Pierre-Robin syndrome and also had a patent ductus arteriosus. He was managed for aspiration pneumonitis and was on oxygen for five days. This child was screened for ROP on the 13th day of life, and was found to have Posterior Zone II, stage 3 plus disease (Type 1 ROP) at the first screening warranting immediate LASER therapy (Fig 1).

Fig 1: Fundus pictures of Stage 3+ROP found on 13th day of life



Case 2

The first of the set of twins was admitted with respiratory distress, hypoglycaemia and sepsis, with a birth weight 1300g. The child required oxygen for nine days, and had persistence of the ductus arteriosus which was closed pharmacologically by day seven of life.

Case 3

The second twin also weighed 1300g and was admitted with respiratory distress and hypoglycaemia. He required oxygen for three days. The twins were screened for ROP for the first time on the 16th day of life, at which time they had ROP in Zone II, early stage 3 disease, and no plus. This progressed to stage 3, pre-plus ROP by the second screening on the 23rd day of life (Figures 2 and 3)

Fig 2: Stage 3 pre plus ROP on 23rd day of life in Twin 1

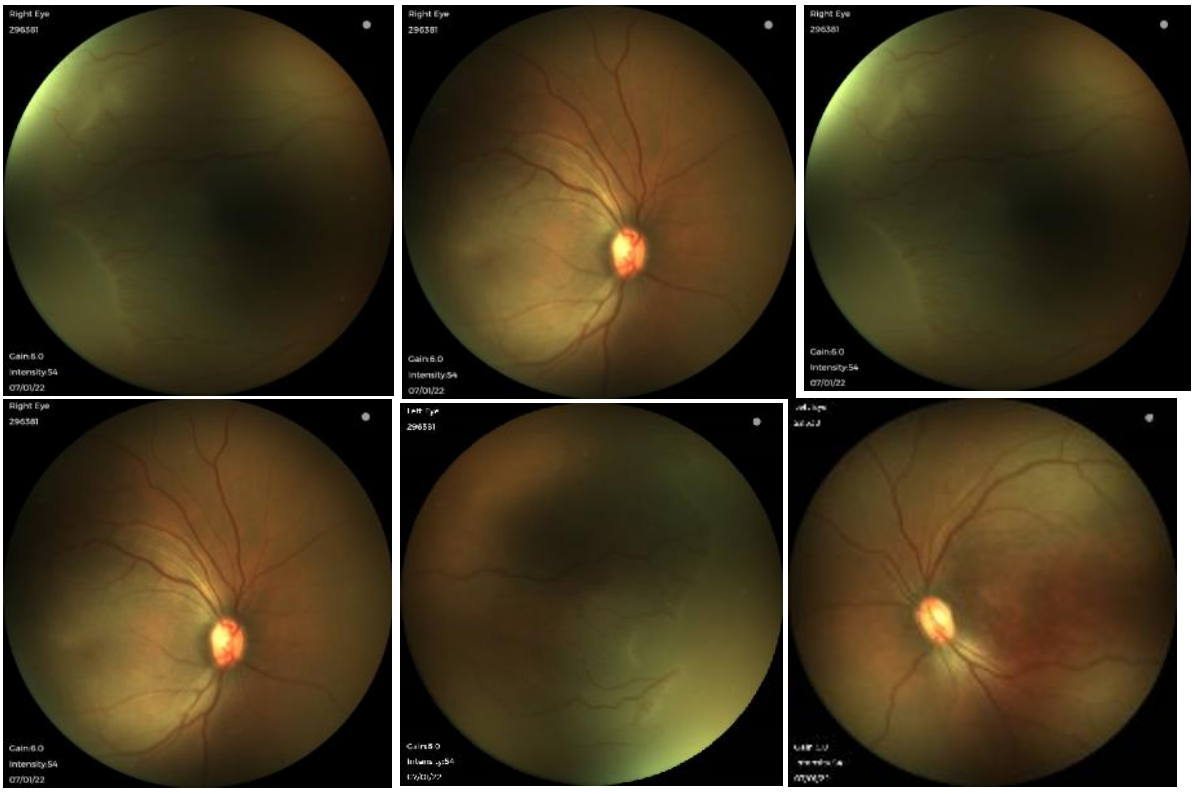
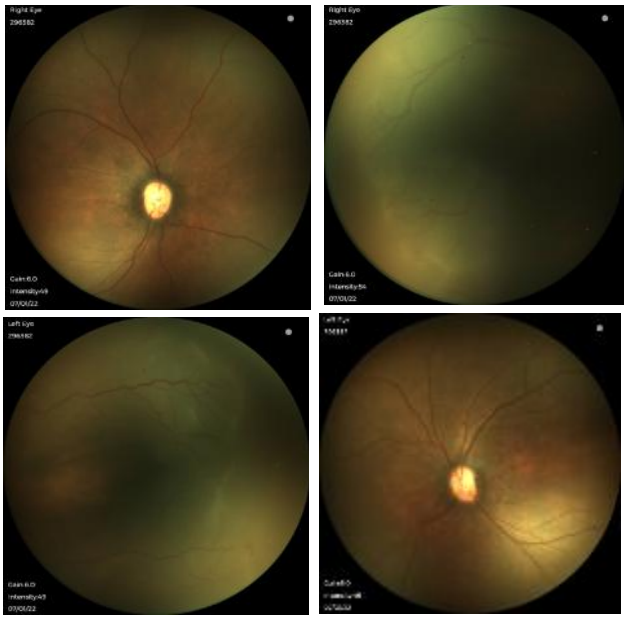


Fig 3: Stage 3 pre plus ROP on 23rd day of life in Twin 2



Discussion

These case reports highlight the fact that with regards to ROP screening criteria, 'one size does not fit all': larger babies can also develop sight threatening ROP within the first two weeks of life. The normal screening protocol for ROP in our hospital ensures that all eligible babies are screened before discharge to establish a relationship with the family so that the babies are not lost to follow up. This protocol enabled detection of potentially sight-threatening ROP in these babies early enough in life for close monitoring and appropriate intervention.
Smith et al in the year 1994 demonstrated mouse model of oxygen induced retinopathy which showed retinal neovascularization between day 17 and day 21 postnatally.[3] Subsequent research on the pathogenesis of ROP elucidated the two phases of ROP development leading up to increased VEGF production and therefore neovascularization. It has been generally accepted that phase 1 occurs from 22 to 32 weeks post menstrual age, and phase 2 from 31 to 44 weeks.[4]The initiation of screening for ROP based on the foregoing and on evidence from the natural history data from the Cryotherapy for Retinopathy of Prematurity study depends on both the postmenstrual age and postnatal age.[5] This occurs at 31weeks post menstrual age in infants with GA of ≤27 weeks at birth, and at four weeks postnatal age in infants with GA >27 weeks at birth.

Some studies have however described a subtype of ROP called the fulminate or rush ROP, also known as aggressive posterior ROP (AP-ROP), which develops earlier than the conventional ROP described by the ICROP classification and which has a different morphology. A 2005 study by Shah et al in Indian babies had babies being screened in the NICU before discharge and subsequent exams were done at the base hospital thereafter. This study had babies who required laser therapy for fulminate ROP as early as 1.5 weeks postnatal age.[6]Babies in developing countries could have earlier AP-ROP, and so it has been recommended that babies less than 28 weeks GA or less than 1200g birth weight should be screened earlier at 2-3 weeks postnatal age to enable early detection and treatment of AP-ROP.[7] The babies in our report had higher birth weight and GA than those required to have earlier screening by what had been previously recommended. Studies have found that intrauterine growth restriction, maternal chorioamnionitis, having culture-proven sepsis, intraventricular hemorrhage and the need for more plasma transfusions, among others, are risk factors for aggressive posterior ROP. [8, 9]One of the babies in this case series had sepsis, while two had persistence of the ductus arteriosus. The role of the congenital heart anomaly, the Pierre Robin syndrome, birth asphyxia, aspiration pneumonitis and even the hypoglycemia found in this report in the earlier onset of ROP warrants further research.

The first epidemic of ROP was brought about by unmonitored oxygen therapy in Europe and America.[10, 11] The third epidemic experienced by developing countries was also due to lack of proper neonatal care and improper oxygen therapy.[12]The babies in this study had five days of oxygen therapy; it has been shown that total days on oxygen therapy is an independent risk factor for development of ROP in preterm babies.[13]
To the best of our knowledge, this is the first description of such early onset ROP among Nigerian babies. It is therefore expedient that babies get screened in the NICU before discharge. There is a need for constant review of ROP screening criteria and timing based on locally available data. Studies have continued worldwide on how best to identify the most at risk babies while reducing the current burden of the number needed to screen to get a case to treat.[14, 15]Further research may be required among Nigerian babies to evaluate the association between the development of type I ROP and number of days on oxygen therapy, using the current system being run in the NICUs. Findings from such research may help guide the need for earlier screening among babies based on the duration of oxygen therapy. It may also point to the need to change the time of screening from four weeks postnatal age to possibly 2-3 weeks of life.

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