

**SUSPECTED CHIASMATIC PATHOLOGY
IN PAZIENT AFFECTED BY ATYPICAL SECTOR
RETINITIS PIGMENTOSA: THE IMPORTANCE
OF CLINICAL AND STRUMENTAL VALUES**

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Introduction

Retinitis pigmentosa (RP) is one of the most common hereditary retinal dystrophies and causes of visual impairment affecting all age groups. Sector retinitis pigmentosa is an atypical form of RP that is characterized by regionalized areas of bone spicule pigmentation, usually in the inferior quadrants of the retina. Typically it presents symmetric and bilateral. Sector RP has a variable symptomatology: night vision decrease (nictalopia), photophobia, visual field constriction, with partial or complete scotoma. However, any pazients may remain asimptomathic until adulthood.

Methods

A 40-year-old Caucasian woman, with a suspected diagnosis

of chiasmal disease, is subjected to a series of exams: manual visual field, Humphrey visual field, elettroretinography (PEV-ERG-EOG), ophthical coherence tomography (OCT), retinical fluoroangiography (FAG), chromatic sense tests (Hishihara and Farnsworth).

Results

With an accurate ophthalmoscopic exam we demonstrated that the patient had an atypical form of retinitis pigmentosa (sectorial) and not a chiasmal disease: bone spicule pigmentation was found in the nasal and inferior quadrants in each eye. We demonstrated also superior temporal visual-field loss corresponding to the areas of the affected retina. Clinical measurements of visual-field loss, best-corrected visual acuity, and ophthalmoscopic appearance have remained stable during the year the patient has been followed.

Discussion

Sector retinitis pigmentosa is an atypical form of RP that is characterized by bilateral pigmentary retinopathy, usually isolated to the inferior quadrants. The remainder of the retina appears clinically normal, although studies have found functional abnormalities in these areas as well. Sector RP is generally considered a stationary to slowly progressive disease, with subnormal electro-retinogram findings and visual-field defects corresponding to the involved retinal sectors. Management of RP is very difficult because there are no proven methods of treatment. For these reasons is very important correlate clinical exams (ophthalmoscopic) with instrumental exams.

Key words: chiasmatic pathology, retinitis pigmentosa.

**NEURODEVELOPMENTAL OUTCOME IN VERY
PRETERM AND TERM INFANTS AT 36 MONTHS
CORRECTED AGE USING THE BAYLEY-III**

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Background

Advances in perinatal care have led to an increase in survival of preterm children but have also increased the risk of long-term sequelae, such as neurodevelopmental impairment and behavioral or emotional disturbance.

Objective

To investigate neurodevelopmental patterns of preterm infants born at S. Maria alle Scotte Hospital, Siena, Italy.

Methods

65 preterm infants born from April 2006 to February 2008 were enrolled.

Inclusion criteria: gestational age (GA) d"32 weeks.

Exclusion criteria: Major congenital malformations, inborn errors of metabolism. Neurodevelopmental follow up was performed at 3 years old, using Bayley scale of Infant and Toddler Development IIIed that consisted of five scales: Cognitive (CS), Language (LS), Motor (MS), Social-Emotional (SES) and Adaptive Behavior Scale (ABS).

Results

The 5 Bayley scores of all subject resulted within the range of normality. IVH was associated with lower motor score (P<0,001). Babies with ROP showed a lower score in 4 Bayley s items (CS: ≤0,01; LS: ≤0,05; MS: ≤0,01; SES: ≤0,05). Multiple regression analysis confirmed as predictors of neurological outcome: GA for cognitive (p=0,016), language (p=0,004) and social-emotional development (p<0,0001), IVH for motor (p<0,0001) and adaptive behavior development (p<0,0001), and twinship for language (p=0,001) and social-emotional development (p=0,003). BPD had a negative effect on cognitive development (P=0,049) while PDA on social-emotional development (p=0,023).

Conclusions

Several well recognized factors has been identified as predictors of poor neurological outcome. Among these factors, BPD and IVH were the best predictors being associated with lower scores at Bayley scales. Surprisingly, twinship appeared to be a protective factor.