

# Improvement of Corneal Opacity in Peters' Anomaly

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## Abstract

A 5-week-old male presented with Peters' anomaly. Corneal transplantation was deferred and the patient underwent an optical sector iridectomy in his better eye with central leukoma and iridocorneal adhesions. At six months following surgery, there was a decrease in corneal opacity. The most recent ophthalmic examination was performed at age 42 months. The cornea was considerably clearer, and the child had a visual acuity of 20/50. Optical sector iridectomy should be considered, in selected cases of Peters' anomaly.

## INTRODUCTION

Peters' anomaly is among the most common causes of congenital corneal opacities requiring keratoplasty. Despite modern surgical techniques, visual outcome after corneal transplantation is less than desirable due to a multitude of postoperative complications such as graft failure, cataract, retinal detachment, and glaucoma (1). Atropinization of the pupil, rotating corneal autograft and optical iridectomy all have been proposed as alternatives, in selected cases.

We performed optical sector iridectomy in a patient with Peters' anomaly and observed a decrease in corneal opacity with excellent visual outcome.

## CASE REPORT

A male child was born 6 weeks prematurely via spontaneous vaginal delivery following an unremarkable pregnancy. He was nursed for 3 weeks in a neonatal unit where photophobia and lacrimation along with cloudiness of his corneas were noted by the nursing staff. Two weeks after discharge, he was examined at the Cornea and External Disease Service of The Wilmer Eye Institute. He had no nystagmus. An examination under general anesthesia revealed total corneal opacity in the right eye (OD). Left eye (OS) showed central corneal opacity with a small clear area superotemporally (Figure 1A). Anterior chamber structures could be visualized through this area; a mildly oval pupil and a clear lens were noted. Corneal diameters were 8.5 x 7.5mm OD and 9.5 x 7.5mm OS. Intraocular pressures measured 12 mmHg OD and 19 mmHg OS. Due to corneal opacities and small pupils, a fundus examination could not be performed.

Ultrasonographic investigations showed a small OD with an axial length of 15.79 mm, shallow anterior chamber, and a large retinal detachment with vitreal band formation. The axial length of OS was 19.37 mm with normal anterior and posterior segment structures.

## Figure 1

Figure 1A: Left eye, pre-operatively, demonstrating a central corneal opacity.



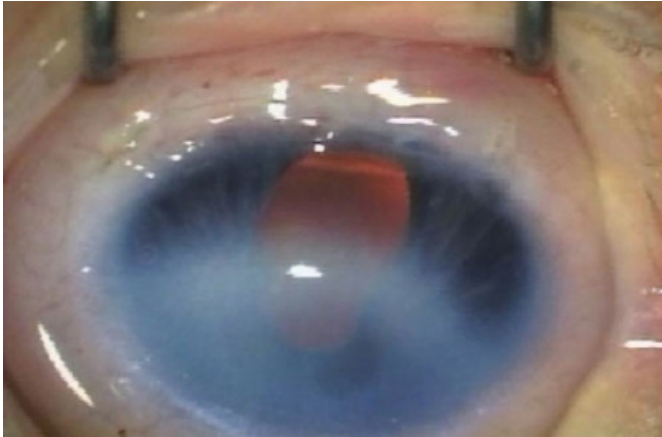
The child was diagnosed with Peters' anomaly. Detailed family history was unremarkable. Available family members were examined and no corneal or other anterior segment abnormalities were found.

In the following 3 weeks, corneal opacity did not change. Thus, a decision to operate the left eye was made. Mechanical separation of iridocorneal adhesions and a large sectoral optical iridectomy was performed at the superior quadrant (Figure 1B). Indirect fundus examination through

the iridectomy, at the completion of the surgery, provided a 60-degree view and demonstrated normal retina, vessels and optic nerve.

### Figure 2

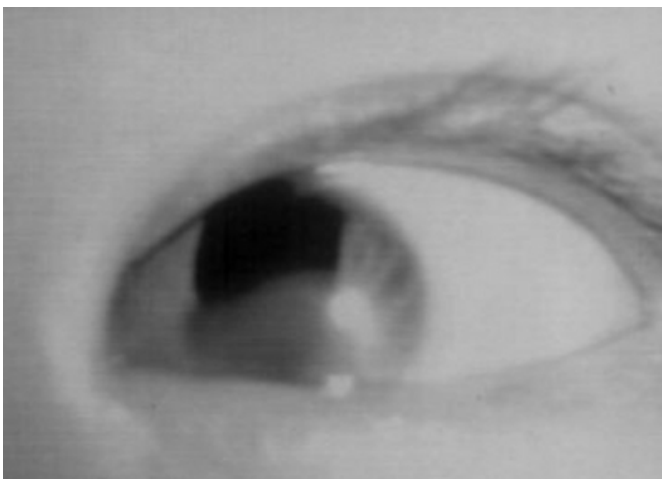
Figure 1B: Left eye, immediate post-operatively, demonstrating a central corneal opacity and sectoral iridectomy.



Six months after surgery a decrease in the corneal opacity was noted (Figure 2). The child was followed closely for an additional 36 months. The latest ophthalmic examination demonstrated a significantly clearer cornea with a visual acuity of 20/50 and normal intraocular pressure.

### Figure 3

Figure 2: The same eye at 6 months post-operatively, with significant clearance of the cornea.



## DISCUSSION

Early visual rehabilitation must be considered in patients with Peters' anomaly, due to the devastating amblyopic effects of dense central leukomas. Penetrating keratoplasty is currently the most commonly performed procedure, with considerably low graft survival rates due to difficulties in

surgical technique as well as high rate of immune rejection. Secondary glaucoma requires multiple additional surgical procedures and is associated with a poorer visual outcome. Unfortunately, only 1/3 of patients maintain a clear graft long-term, with varying functional visual outcomes (2). Earlier work suggested postponing surgery until the patient is about 1-year-old (3).

Optical sector iridectomy was recommended as a conservative surgical approach, alternative to corneal transplantation, with comparable visual results and significantly less post-operative complications (4). In present case with Peters' anomaly, we performed sector iridectomy. The cornea cleared considerably and the patient gained excellent visual acuity. Peters' anomaly of the cornea is likely to result from a faulty migration of the first wave of neural crest cells (5), with a discontinuous endothelial layer. Descemet's membrane and endothelium are present beneath the area of the corneal opacity except at the points of iris adhesion (6). In our case, it is not possible to determine whether the corneal clearing was spontaneous or facilitated by surgical intervention. However, we hypothesize that removal of iridocorneal adhesions might have helped in the migration of the remaining normal endothelial cells to form a somewhat continuous layer.

## SUMMARY

Visual iridectomy in Peters' anomaly seems to be the most adequate approach in cases with a large clear corneal area enabling the performance of a functional visual iridectomy. Optical sector iridectomy should be considered, in selected cases of Peters' anomaly.

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