

# Case Study

Pennsylvania College of Optometry  
Master of Science in Clinical Optometry  
German Course 2004

**Case Number One:  
Fitting of semi-scleral contact lenses in a patient with simple microphthalmos and keratoconus**

by  
Raymond E. Waelti  
Optilens Kontaktlinsen  
Baelliz 67  
CH-3601 Thun  
Switzerland  
Phone: +41 33 222 54 22  
Fax: +41 33 222 57 22  
Email: raymond.waelti@optilens.ch

**Abstract:**

Microphthalmos is a congenital and mostly hereditary condition of one or both eyes. Other ocular abnormalities also occur, including cataract, glaucoma, aniridia, and coloboma. Systemic and anatomic abnormalities also often occur<sup>1</sup>. Keratokonius is a progressive disorder in which the cornea assumes an irregular conical shape. The hallmark of keratokonius is central or paracentral stromal thinning, apical protrusion and irregular astigmatism. The onset is around puberty and progresses slowly thereafter, although it may become stationary at any time. The role of heredity has not been clearly defined and most patients do not have a positive family history<sup>2</sup>. This case report shows a special way of CL fitting in a keratokonius patient with microphthalmos and discusses the risks and benefits thereof.

**Key words:** *microphthalmos, keratokonius, semi-scleral contactlens*

## Introduction

Microphthalmos is the term to describe an eye that is smaller than normal. Microphthalmos is the general term which is subdivided in pure microphthalmus (nanophthalmus), simple microphthalmus and complex microphthalmus<sup>3,4</sup>.

Pure microphthalmos also called nanophthalmos describes an eye that is smaller than normal but with a normal size lens. Usually both eyes are affected. One thinks that the condition occurs because the globe stops growing at a certain point of time during the development. The size of the lens is as in a normal eye. This is the reason why nanophthalmic eyes have shallow anterior chambers and are highly hyperopic. Therefore there is an increased risk of angle-closure-glaucoma<sup>3,4</sup>.

Simple microphthalmos is different from pure microphthalmos by the fact that the lens size is also reduced. Therefore the eye is not highly hyperopic and no increased risk of angle-closure-glaucoma is existent. Most cases are sporadic and bilateral. It represents a retardation in eye growth after the primary optic vesicle has formed and invaginated. Simple microphthalmos may be caused by radiation, mechanical stimulation and chemicals. It can also result from intrauterine toxoplasmosis, rubella or as a component of a ocular-genetic syndrome<sup>3,4</sup>.

Complex microphthalmos is describing a small deformed eye. It can be unilateral or bilateral and include several additional ocular deformations such as anterior segment maldevelopments, hyperplastic primary vitreous and other lens, vitreous or retinal abnormalities. Microphthalmos with coloboma is a specific type of complex microphthalmus and can be associated with heart defects<sup>3,4</sup>.

Keratoconus is a bilateral, asymmetric, cone-shaped deformity of the cornea. The underlying process is a progressive thinning of the paracentral cornea. Keratoconus is often associated with systemic disorders such as: Down Syndrome, Turner Syndrome, Ehlers-Danlos Syndrome, Marfan Syndrome, atopy, osteogenesis imperfecta and mitral valve prolaps. Also associations with other ocular diseases are common such as: vernal disease, Leber congenital amaurosis, retinitis pigmentosa, blue sclera, aniridia and ectopia lentis. In addition to the above mentioned possible predisposing factors are also PMMA contactlens wear and constant eye rubbing<sup>2,5</sup>.

According to Amsler the Keratoconus can be divided into four grades corresponding the signs visible at the cornea<sup>6</sup>.

- Grade1: regular mixed or myopic astigmatism forms. Ophthalmometer mires slightly displaced. Good Visual Acuity (VA) with glasses.
- Grade2: Shorter corneal radii, and higher astigmatism. Still relatively good VA with glasses and no remarkable structures visible with the slitlamp. Often ophthalmoscopy shows a "oil droplet" reflex.
- Grade3: Cone-like deformation of the cornea visible with the slit-lamp. Only poor VA can be achieved with glasses. Slit-lamp biomicroscopy shows very fine, deep, stromal striae (Vogt lines)
- Grade4: Increased deformation and paracentral thinning of the cornea. Munson sign (bulging of lower lids on downward gaze), corneal scarring, Fleischer ring (epithelial iron deposits at base of the cone), descemet folds and prominent corneal nerves may be present. Acute hydrops can develop.

## Case Report

At 21 October 2000, H.P., a 39 year old white male (office worker) presented to our contact lens practice with a complaint of severe photophobia and foreign body sensation while wearing his rigid-gas permeable (RGP) lenses. The condition affected both eyes and existed since several months. His former contact lens practitioner refitted him with new contacts which did not eliminate the symptoms. The patient was told to continue to wear his contacts to get used to them.

He had a 12 year history of comfortable RGP lens wear with no incidents of infections or inflammations. His last exam with an ophthalmologist was in September 2000, about one month prior to his visit at our institute. Twelve years ago he was diagnosed with keratoconus OU. He has no systemic disease and no family history of any eye- or systemic disease. He was oriented to time, place and person.

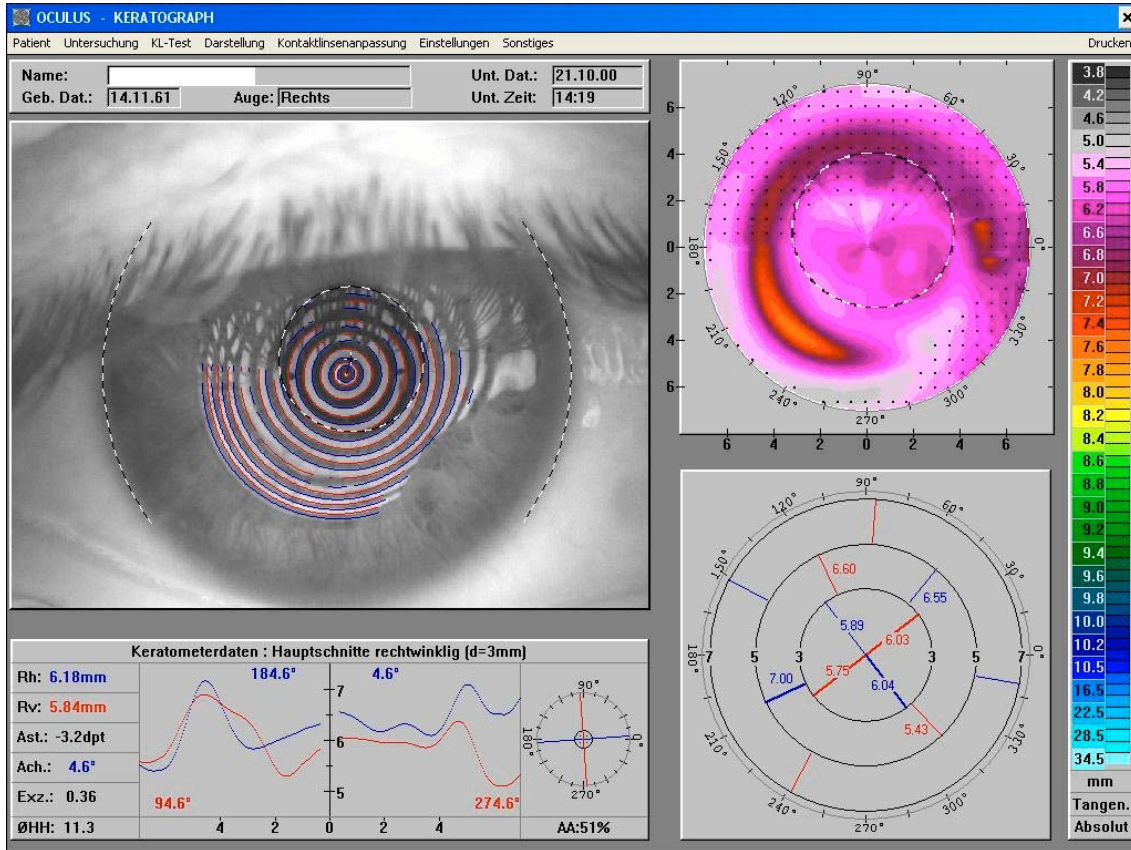
His uncorrected visual acuity was 20/400 at distance OD, and 20/400 at distance OS after removal of his current RGP lenses. Uncorrected near acuities were: 20/16 (.3M) @ 3.5cm OU. Best-corrected visual acuity with glasses was 20/50 OD and 20/50 OS. The refraction upon removal of his RGP lenses was  $-11.75 -4.00 @180^\circ$  OD and  $-11.00 -8.00 @180^\circ$  OS at a corneal apex to lens distance of 14mm OU. Pinhole acuity at distance was 20/100 OD and 20/100 OS. Color vision testing with pseudoisochromatic plates showed no color vision deficiency OU. Pupils were equally round and reactive to light, no afferent pupil defect was noted OU. Confrontation fields were full to finger count OU. Extraocular muscles were unrestricted in all gazes, and cover test demonstrated orthophoria at distance and near. Due to the law of practice in Switzerland no Goldman Tonometry could be performed. Anterior segment evaluation by slit lamp examination revealed a quiet bulbar and palpebral conjunctiva OU. An even tear film with tear break up time of 15 seconds OU. Clear lashes OU with a bilateral, symmetrical ptosis. The patient reported no change in the eyelid position since he remembered. Due to the symmetrical character, the presence of a lidfold OU and the patient's report the ptosis was noted to be congenital. Also the globes presented exceptionally small OU. The horizontal diameter of the cornea was noted 11.30mm OD and 11.10mm OS. The lens appeared to be in normal dimension compared to the size of the globes and no hyperopia or shallow anterior chamber was noted. Therefore the diagnosis of simple microphthalmos was made. The corneas showed the following: Vogt lines, Fleischer ring and prominent nerves OU. Temporal fluorescein staining due to CL wear OU. No Descemet folds or scarring was noted and the corneas were clear OU. Iridides were brown OU. The anterior chamber appeared clear without cells or flare and the anterior chamber angles were estimated by the Van Herrick method with the slit-lamp as 4 nasally and 4 temporally OU. Both lenses were evaluated by slit-lamp with undilated pupils and have been noted as clear with no opacities in any region.

The evaluation of the posterior segment by slit lamp with 90D lens and undilated pupils revealed normal optic nerves with a cup-to-disc ratio of .3/.3 OU. The neuroretinal rims were healthy and intact. Retinal vessels appeared normal with an arterial-venous ratio of 2/3 OU. Both eyes presented with clear maculas without foveal reflex.

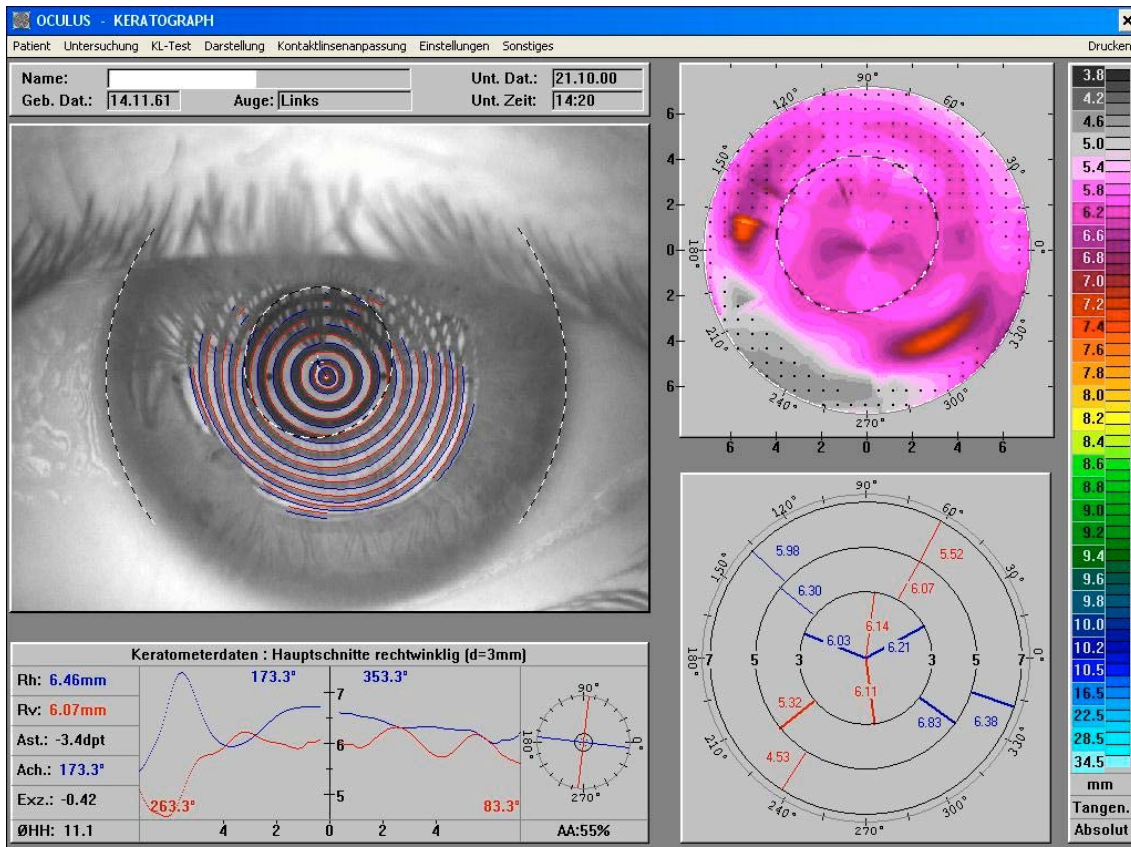
The patient was at this time wearing RGP lenses with the following parameters: OD: KA3 (3-curved back surface) 6.45 (52.37)  $-9.50$  9.20, OS: KA3 6.60 (51.12)  $-9.50$  8.20, both lenses in Boston 4 Material manufactured and fitted in September 2000. The fitting of both lenses was noted as central and midperipheral steep and peripheral flat fit. The visual acuity was noted as 20/20 OD and 20/25 OS.

Due to the fitting pattern of the lenses the Keratometry data was altered. Nevertheless Keratography was performed and revealed the following data:

OD:



OS:



**Differential Diagnosis for this case includes the following:**

- Corneal Dystrophy
- Keratoconus
- Keratoglobus
- Contactlens related corneal distortion
- Contactlens related ocular infection
- Microcornea
- Microphthalmos

Corneal dystrophies are a group of progressive, usually bilateral and mostly genetically determined corneal opacifying disorders which develop in the absence of inflammation. The age at presentation varies between the first and fourth decade depending on the relative frequency of secondary recurrent erosions and on the rate of visual loss<sup>2</sup>.

The signs of Keratoconus have already been described above.

Keratoglobus is a rare, globular deformity of the cornea due to diffuse thinning that is maximal at the base of the protrusion<sup>5</sup>.

Contactlens related distortion of the cornea occurs in a mal-fitted RGP wearer. It resolves spontaneously in a period of four to eight weeks after discontinuing contactlens wear.

Contactlens related ocular infection results from bad contactlens care or infected lenses. It can affect different structures of the anterior globe.

Microcornea is defined as to be an adult cornea smaller than 10mm in horizontal diameter. It is a very rare, hereditary uni- or bilater condition<sup>2</sup>.

Microphthalmos has already been described in detail above.

Due to the altered corneal surface from the worn RGP lenses it was decided to discontinue the RGP lenswear. To enable the patient to continue his daily business custom made soft lenses were fitted to be worn for four weeks.

The patient was scheduled for lens trial seven days later.

**Follow-Up #1, 28 October 2000**

The data of the soft lenses was as follows:

Swisslens, TorisL Igel77 (dynamic stabilized soft lens with 77% water content)

OD: 8.20 -10.00 -2.75 @170° 14.00

OS: 8.20 -9.25 -5.12 @180° 14.00

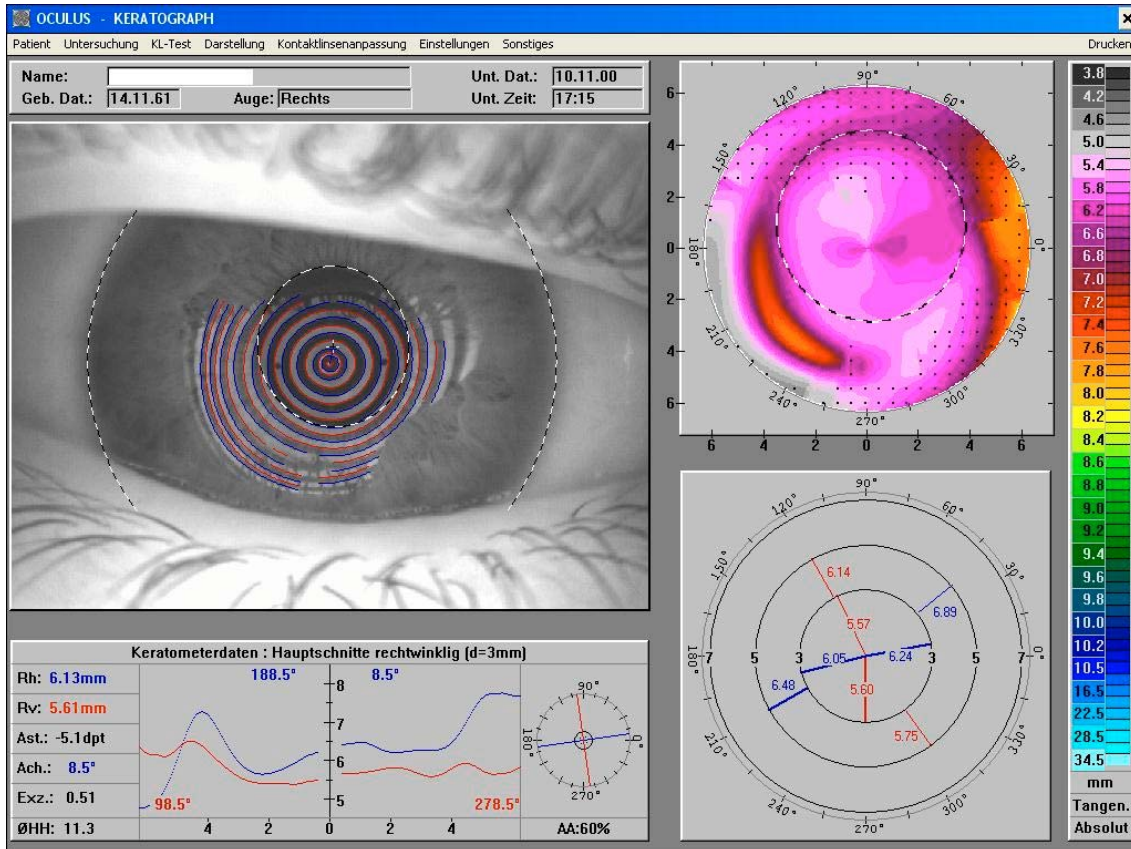
The lens fit was steep and partially bubbles were trapped underneath the lens. The binocular VA was noted as 20/25 which was sufficient for daily living at the time.

**Follow-Up #2, 10 November 2000**

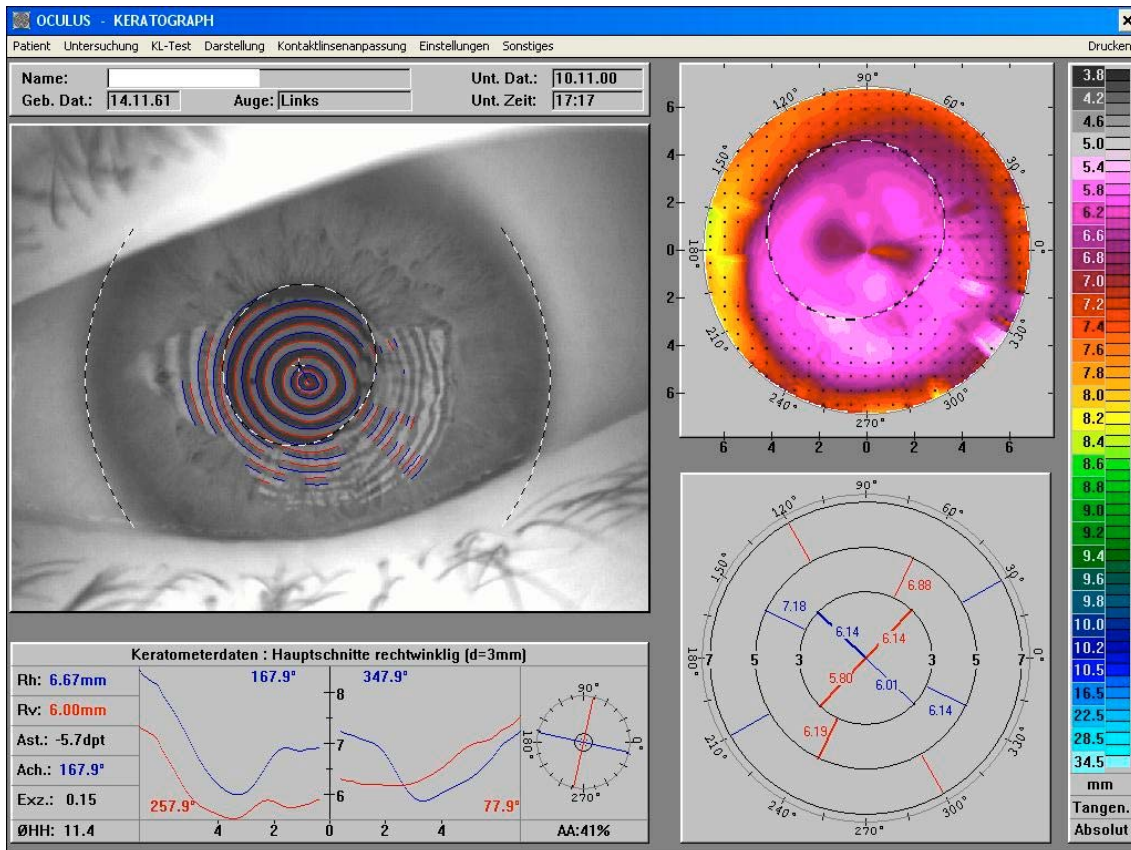
Keratography was performed and RGP trial lens trial performed.

The result of the Keratography was as follows:

OD:



OS:



The following trial lenses were inserted:

OD: Falco, FKK2 5.90 –12.37 9.30

OS: Falco, FKK2 6.00 –11.87 9.30

The over-refraction was noted as:

OD: -2.50 –2.25 @90° VAcc 20/22

OS: -2.50 –2.25 @180° VAcc 20/25

Custom made semiscleral RGP lenses were ordered and a new appointment with the patient was scheduled.

### **Follow-Up #3, 18 november 2000**

The following lenses were fit:

Falco, SKKP (RGP semiscleral prismatic lens with .8 excentricity) Boston XO no tint

OD: 6.00 –15.12 12.00 .8e

OS: 6.10 –15.12 12.00 .8e

The fitting seemed to be steep OU and VA could not be taken due to tearing. The patient was instructed in the handling of the lenses and lens care regimen was explained to him. He was advised to start wearing the lenses one hour the first day and expand the wearing time one hour per day.

### **Follow-Up #4, 24 november 2000**

The patient felt extremely comfortable with the lenses and therefore already was wearing the lenses during the whole waking hours. The foreign-body sensation was no longer present and the patient had no incidents of photophobia since wearing the new lenses. Nevertheless the reduced visual acuity due to a still present astigmatism was not well tolerated.

The fitting was noted to be parallel in the center of the cornea with no touch to the apex. It fitted flat in the mid-periphery and again parallel in the limbal region and on the conjunctiva. Due to the bridging of the lens in the mid-periphery bubbles of air have been trapped underneath the lens. The stabilisation of the lens was noted @20° OD and @170° OS

An overrefraction was performed:

OD: +1.75 –1.50 @140° VAcc 20/20

OS: +2.25 –1.75 @30° VAcc 20/22

Slit-lamp findings revealed a clear cornea without staining and a non-hyperaemic Conjunctiva. The surface of the lenses was well wetted. Also tear exchange underneath the lens was checked with NaFl and found to be adequate.

The lenses were returned to the manufacturer for the changes in power to be made and to drill ventilation holes to allow the air bubbles to escape.

### **Follow-Up #5, 9 december 2000**

The following lenses were dispensed:

Falco, SKKPTN Boston XO, no tint

OD: 6.00 –13.37 –1.25 120° 12.00 .8e

OS: 6.10 –13.12 –1.25 40° 12.00 .8e

The fitting was corresponding to the findings of the last exam. The visual acuities were noted as 20/20 OD and 20/22 OS.

The patient was advised to wear the lenses during the whole waking period and to return for checkup.

### **Follow-Up #6, 13 december 2000**

No changes were noted in the fitting or visual acuity. Also slitlamp findings corresponded with the findings of the exam on 24 november 2000.

The patient was advised to continue lenswear during the whole waking period and to report any problems such as photophobia, tearing, foreign body sensation or lens adherence. The next exam was scheduled one month later.

### **Follow-Up #7 17 january 2001**

The patient reported no incidence photophobia or foreign body sensation. The removal of the lens was reported to be difficult at times due to lens adherence. He was happy with his visual acuities, comfort and wearing time of the lenses.

The slitlamp evaluation revealed a clear cornea OU with grade 1 central staining OD and a grade 2+ central staining OS. The conjunctiva was without hyperemia OU.

Refraction was performed and yielded the following information: OD: plano VAcc 20/20, OS: plano VAcc 20/22.

I was concerned about the staining especially of the left eye and therefore decided to modify the left lens. A new lens was custom made with a larger excentricity to prevent lens adherence.

### **Follow-Up #8 31 january 2001**

Dispensing of the new lens OS with following parameters:

Falco, SKKPT, Boston XO, no tint

OS: 6.10 -13.00 -1.12 @40° 12.00 .9e

The slitlamp examination of this day revealed a clear cornea without staining OU even though the former lenses have still been worn.

Rechecks were performed at the following dates:

21 february 2001

31 may 2001

31 july 2001

All exams showed a clear cornea without staining OU and a quiet conjunctiva OU. Visual acuities were stable at the mentioned limits. The new lens OS was not well tolerated by the patient due to more lens movement. After the first few days he therefore wore the former lens OS, again. Due to a better method of removal of the lens no more corneal staining was noted during all of the exams.

The patient was re-instructed in the lens care and wearing schedule and a semi-annual re-check interval was proposed.

### **Discussion**

Keratoconus is in most cases a progressive disease. In the early stages glasses can help to reach a good visual acuity for daily living. Since the presentation of a Keratoconus includes a progressively irregular shaped cornea the fitting of a contactlens is inevitable at a certain point of time. The best visual acuity can be reached with a RGP lens. Additional circumstances can enhance the difficulties of a lens fitting. For example Microphthalmos and Ptosis. Both force the lens-fitter to rely on custom made lens designs to ensure proper lens fitting with minimal side effects on the eyes physiology.

## Conclusion

A combination of Keratoconus and Microphthalmos presents a challenge for the contactlens specialist. Especially if the patient is sensitive to RGP contacts.

In some cases a large, semiscleral contactlens can be of great help. The advantages are a very good spontaneous compatibility and a good optical quality. Another advantage can be the longevity of a RGP lens. I had the best results in replacing large RGP lenses at least every two years.

The risks of a large RGP-fitting include lens adherence with deformation and or staining of the cornea. This enhances the risk of infection. A strict lens care regimen and close check-up intervals plus the replacement of the lenses at least every two years can help to minimize these risks.

## Bibliography

1. Definition by :Texas School of the blind and visually impaired ([www.tsbvi.edu](http://www.tsbvi.edu))
2. Clinical Ophthalmology, Jack J. Kanski, Fourth Edition, 1999
3. Definition and explanation by [www.cougarhealth.com](http://www.cougarhealth.com)
4. Schwierigkeiten und Komplikationen der Cataract-Chirurgie mit Linsenimplantation bei Mikrophthalmus anterior, H.-E. Voelcker, Universitaets-Augenklinik Heidelberg, Heidelberg. [www.dgii.org/1996/ref10.html](http://www.dgii.org/1996/ref10.html)
5. The Massachusetts Eye and Ear Infirmary Illustrated Manual of Ophthalmology, 1998
6. Augensymptome, Joerg Trotter,Optik-Verlag, Trimbach, Switzerland, 1992