

## Keratoconus in Civil Aviation Pilots in a Report of Six Cases

Albert Rebello; Bênesson Rodrigues; Marcos Pereira

- BACKGROUND:** Keratoconus is a progressive noninflammatory ectatic corneal dystrophy, characterized by corneal thinning and increased curvature with central or paracentral tapered protrusion. Such changes in corneal morphology induce irregular astigmatism and myopia, resulting in visual acuity impairment. This report assesses cases in which keratoconus is not an absolute disabling condition for civil aviation pilots.
- METHODS:** Six cases of keratoconus in active pilots are presented. This report includes cases which were treated with crosslinking therapy and even corneal transplantation. In such cases, the civil aviation medical authority considered four aspects in the medical assessment: best corrected visual acuity, condition stability, risk of sudden incapacitation, and pilot category.
- RESULTS:** Six civil pilots with ages between 19 and 45 yr of age presented with keratoconus in different stages. Using the Snellen scale, best corrected visual acuity (BCVA) was measured in 12 eyes, all of them having equal or better than 20/30 (20/20,  $N = 9$ ; 20/25,  $N = 2$ ; 20/30,  $N = 1$ ). All of them are currently fit for aviation activity and have been given an aviation medical certificate with limitation of use of corrective lenses by the Brazilian Civil Aviation Authority (ANAC in Portuguese). Among these cases, there is a pilot who had undergone treatment with crosslinking therapy and another that had undergone corneal transplantation.
- DISCUSSION:** These cases demonstrate that keratoconus is not always a disabling condition for civil aviation pilots. Nevertheless, it has to be analyzed on a case-by-case basis.
- KEYWORDS:** keratoconus, civil aviation, corneal pathology.

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Keratoconus is a progressive noninflammatory ectatic corneal dystrophy, characterized by corneal thinning and increased curvature with central or paracentral tapered protrusion.<sup>6,9</sup> Such changes in corneal morphology induce irregular astigmatism and myopia, resulting in visual acuity impairment.<sup>7,9</sup> The disease occurs in all races, bilaterally and asymmetrically, with an incidence of 1:2000.<sup>1</sup> It presents a progressive visual impairment, although is more stabilized after the fourth decade. Symptoms are highly variable and, in part, depend on the stage of the progression of the disorder. Early in the disease, keratoconus may be noted by the ophthalmologist simply because the patient cannot be refracted to a clear 20/20 corrected vision. In advanced disease, there is significant distortion of vision accompanied by profound visual loss.<sup>9</sup> Depending on the severity of the disease, the followings signs may be detected by slit-lamp examination: stromal corneal thinning, conical protrusion, iron line surrounding the cone, fine vertical lines in the deep stroma and Descemet's membrane

(Vogt's striae), and stromal scars. In patients with advanced disease, the risk of sudden incapacitation with acute hydrops (painful corneal edema) is augmented if there are previous corneal scars and corneal thinning. Usually, hydrops occurs in one eye separately, but it is sufficient to incapacitate the subject due to the painful manifestation.

The initial visual rehabilitation is performed with the use of glasses. However, there are cases where the refraction shows high and irregular astigmatism. In these cases, whenever

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rehabilitation is not achieved with the use of special contact lenses, surgical techniques become necessary.

Currently, new techniques have been used, such as intracorneal implant ring or covalent crosslinking promotion of the corneal collagen (crosslinking) by stimulation with riboflavin and ultraviolet radiation. These techniques are able to delay or even avoid a traditional treatment with corneal transplantation, indicated whenever intolerance to contact lenses use is found.

In military aviation, the pathology is an absolute disabling condition. But in civil aviation, the development of new therapies currently allows assessing an applicant's fitness using the flexibility standard in some cases. Such flexibility has been noticed in different regulations of international civil aviation authorities, such as the Federal Aviation Administration (FAA, USA),<sup>5</sup> the European Aviation Safety Agency (EASA),<sup>8</sup> the Civil Aviation Authority (CAA, England),<sup>10</sup> and the Civil Aviation Safety Authority (CASA, Australia).<sup>4</sup>

## METHODS

### Subjects

This manuscript is a summarized presentation of six individual cases of currently flying aviators. It is not a research project, but just a series of six case reports. All subjects had authorized sending medical information to the Brazilian Civil Aviation Authority. The identities of the subjects were protected by the Brazilian Civil Aviation Authority, as specified in Annex 1 of the International Civil Aviation Convention: "1.2.4.10.1. All medical reports and records shall be securely held with accessibility restricted to authorized personnel."

There is an exemption from approval by committee for ethical review of human experimentation. The Brazilian medical aviation authority does not act as pilots' treating physician. Like other international civil aviation authorities, our medical doctors receive reports from pilots' treating physicians and analyze how the medical condition could be assessed as not likely to interfere with the safe exercise of the pilot's license or rating privileges.

Six civil aviation pilots between the ages of 19 and 45 yr presented with keratoconus in different stages. All subjects are Caucasian and of the male gender.

### Equipment

Concerning to the six cases here described, best corrected visual acuity (BCVA) at distance was measured by the Snellen chart, where the view scale level 20/20 corresponds to visual acuity 1.0 in the decimal scale. The corneal thickness, when measured, was verified by ultrasonic pachymetry. Measurement lower than 450 microns was considered below the average population.<sup>11</sup> Corneal curvature measure (keratometry) was evaluated with computerized corneal topography. For comparison purposes, keratometry was taken in the central cornea (3 mm diameter), as well as measurement of the greater corneal curvature.

### Procedure

To mitigate the risk of poor visual performance, an operational limitation had been imposed on applicants who had not achieved BCVA 20/20 in each eye. In these cases, pilots could only fly with another pilot (safety pilot).

When it was verified that an applicant had undergone a surgical procedure, medical assessors of the civil aviation authority requested a report from the treating physician about refraction stability. Accordingly with this report, a reduced validity was applied. In all cases, candidates were released to fly using corrective lenses.

### Case 1

In 2015, a 24-yr-old Caucasian male applicant had undergone a medical assessment for airline pilot licensing. Ophthalmological examination showed uncorrected visual acuity in the right eye (OD) at 20/25 and in the left eye (OS) at 20/40. With use of corrective lenses, he has achieved a BCVA in OD of 20/25 and OS of 20/25. No corneal scars, corneal opacities, or Vogt's striae were shown on ocular biomicroscopy (slit-lamp examination). No significant data were shown on ocular motility exam, intraocular pressure measure, or fundoscopy. The corneal topography data and pachymetry ultrasonic of the cornea are presented in **Table I**. Keratometry in the right eye was 44.1 × 48.4 D, with a greater curvature of 54.1 D, and in the left eye was 42.4 × 46.6, with a greater curvature of 52.8 D. Corneal thickness in OD was 482 microns and in OS was 495 microns.

The patient had a history of keratoconus treatment with crosslinking in 2013 in both eyes. Considering possible changes in refractive errors in this case, the medical assessors of the Brazilian Civil Aviation Authority (ANAC) authorized the issuance of the medical certificate with validity restriction, requesting medical evaluation on a semester basis. An operational limitation has been imposed too. In this case, the pilot could only fly with another pilot (safety pilot), as shown in **Table II**.

### Case 2

In 2015, a 45-yr-old Caucasian male applicant had undergone a medical assessment for airline pilot licensing. Ophthalmological examination showed uncorrected visual acuity in OD of 20/30 and in OS of 20/30. With use of corrective lenses, he has achieved a BCVA in OD of 20/20 and in OS of 20/20. No corneal scars, corneal opacities, or Vogt's striae were shown on ocular biomicroscopy (slit-lamp examination). No significant data were shown on ocular motility, intraocular pressure, or fundoscopy. The corneal topography data and pachymetry ultrasonic of the cornea are presented in **Table I**. Keratometry in the right eye was 47.8 × 44.9 D, with a greater curvature of 51.8 D, and in the left eye was 46.6 × 45.0, with a greater curvature of 50.5 D. Corneal thickness in OD was 474 microns and in OS was 438 microns. A medical certificate was issued with validity restriction, requesting medical evaluation every 6 mo. No operational limitation has been imposed.

**Table I.** Corneal Topography and Pachymetry Ultrasonic Data.

| CATEGORY                    | BEST UNCORRECTED VISUAL ACUITY | BEST CORRECTED VISUAL ACUITY | KERATOMETRY (D) GREATER CURVATURE | KERATOMETRY (3 mm)                 | PACHIMETRY                         |
|-----------------------------|--------------------------------|------------------------------|-----------------------------------|------------------------------------|------------------------------------|
| ALP*                        | OD: 20/25<br>OS: 20/40         | OD: 20/25<br>OS: 20/25       | OD: 54.1<br>OS: 52.8              | OD: 44.1 × 48.4<br>OS: 42.4 × 46.6 | OD: 482 microns<br>OS: 495 microns |
| ALP                         | OD: 20/30<br>OS: 20/30         | OD: 20/20<br>OS: 20/20       | OD: 51.8<br>OS: 50.5              | OD: 47.8 × 44.9<br>OS: 46.6 × 45.0 | OD: 474 microns<br>OS: 438 microns |
| CP**                        | OD: 20/300<br>OS: 20/60        | OD: 20/30<br>OS: 20/20       | OD: 48.7<br>OS: 45.3              | OD: 46.8 × 42.5<br>OS: 42.6 × 41.7 | ---                                |
| PP***—2 <sup>nd</sup> class | OD: 20/200<br>OS: 20/200       | OD: 20/20<br>OS: 20/20       | OD: 53.2<br>OS: 49.4              | OD: 48.7 × 46.9<br>OS: 48.9 × 48.5 | ---                                |
| PP—2 <sup>nd</sup> class    | OD: 20/100<br>OS: 20/40        | OD: 20/20<br>OS: 20/20       | OD: 52.6<br>OS: 47.3              | OD: 45.6 × 50.3<br>OS: 45.8 × 46.9 | OD: 499 microns<br>OS: 517 microns |
| PP—2 <sup>nd</sup> class    | OD: 20/60<br>OS: 20/200        | OD: 20/20<br>OS: 20/20       | OD: 48.9<br>OS: 46.5              | OD: 41.8 × 45.5<br>OS: 41.8 × 44.4 | OD: 507 microns<br>OS: 511 microns |

\* ALP – airline pilot ; \*\*CP – commercial pilot; \*\*\*PP – private pilot.

### Case 3

In 2015, a 42-yr-old Caucasian male applicant had undergone a medical assessment for commercial pilot licensing. Ophthalmological examination showed uncorrected visual acuity in OD of 20/300 and in OS of 20/60. With use of corrective lenses, he has achieved a BCVA in OD of 20/30 and in OS of 20/20. No corneal scars, corneal opacities, or Vogt's striae were shown on ocular biomicroscopy (slit-lamp examination). No significant data were shown on ocular motility, intraocular pressure, or funduscopy. The corneal topography data are shown in Table I. Keratometry in the right eye was 46.8 × 42.5 D, with a greater curvature of 48.7 D, and in the left eye was 42.6 × 41.7, with a greater curvature of 45.3 D. There were no corneal pachymetry data.

Considering refraction stability, a medical certificate was issued with validity restriction, requesting medical evaluation on a yearly basis. An operational limitation was imposed due to considerations of BCVA, pilot category, and age. In this case, the pilot could only fly with another pilot (safety pilot limitation).

### Case 4

In 2015, a 35-yr-old Caucasian male applicant had undergone a medical assessment for private pilot licensing. Ophthalmological examination showed uncorrected visual acuity in both

eyes of 20/200. With use of corrective lenses, he has achieved BCVA in OD of 20/20 and in OS of 20/20.

The patient has a history of keratoconus treatment with corneal transplantation in 2001 in the right eye. On slit-lamp examination (corneal biomicroscopy), there was only a peripheral corneal scar caused by the previous eye surgery, sparing the central optical area in the right eye. No other corneal scars, corneal opacities, or Vogt's striae were shown in either eye. No significant data were shown in the ocular motility exam, intraocular pressure measure, or funduscopy.

The corneal topography data are shown in Table I. Keratometry in the right eye was 48.7 × 46.9 D, with a greater curvature of 53.2 D, and in the left eye was 48.9 × 48.5, with a greater curvature of 49.4 D. There were no corneal pachymetry data.

A medical certificate was issued with validity restriction, requesting medical evaluation on a yearly basis. No other operational limitation has been imposed.

### Case 5

A 35-yr-old Caucasian male applicant had undergone a medical assessment for private pilot licensing. Ophthalmological examination showed uncorrected visual acuity in OD of 20/100 and in OS of 20/40. With use of corrective lenses, he has achieved a BCVA in OD of 20/20 and in OS of 20/20. No

**Table II.** Aeromedical Disposition of Each Case.

| CATEGORY                    | AGE | BEST UNCORRECTED VISUAL ACUITY | BEST CORRECTED VISUAL ACUITY | VALIDITY | LIMITATION | PREVIOUS TREATMENT               |
|-----------------------------|-----|--------------------------------|------------------------------|----------|------------|----------------------------------|
| ALP*                        | 24  | OD: 20/25<br>OS: 20/40         | OD: 20/25<br>OS: 20/25       | 6 mo     | PVS-PVI    | Crosslinking, both eyes/2013     |
| ALP                         | 45  | OD: 20/30<br>OS: 20/30         | OD: 20/20<br>OS: 20/20       | 6 mo     | ---        | ---                              |
| CP**                        | 42  | OD: 20/300<br>OS: 20/60        | OD: 20/30<br>OS: 20/20       | 12 mo    | PVS-PVI    | ---                              |
| PP***—2 <sup>nd</sup> class | 35  | OD: 20/200<br>OS: 20/200       | OD: 20/20<br>OS: 20/20       | 12 mo    | ---        | Corneal transplantation, OD/2001 |
| PP—2 <sup>nd</sup> class    | 35  | OD: 20/100<br>OS: 20/40        | OD: 20/20<br>OS: 20/20       | 12 mo    | ---        | ---                              |
| PP—2 <sup>nd</sup> class    | 19  | OD: 20/60<br>OS: 20/200        | OD: 20/20<br>OS: 20/20       | 12 mo    | ---        | ---                              |

\* ALP – airline pilot ; \*\*CP – commercial pilot; \*\*\*PP – private pilot; PVS: safety pilot limitation; PVI: instructing flight limitation.

corneal scars, corneal opacities, or Vogt's striae were shown on ocular biomicroscopy (slit-lamp examination). No significant data were shown on ocular motility, intraocular pressure, or funduscopy. The corneal topography and ultrasonic pachymetry data are shown in Table I. Keratometry in the right eye was 45.6 × 50.3 D, with a greater curvature of 52.6 D, and in the left eye was 45.8 × 46.9, with a greater curvature of 47.3 D. Corneal thickness in OD was 499 microns and in OS was 517 microns.

A medical certificate was issued with validity restriction, requesting medical evaluation on a yearly basis. No other operational limitation has been imposed.

### Case 6

A 19-yr-old Caucasian male applicant had undergone a medical assessment for private pilot licensing. Ophthalmological examination showed uncorrected visual acuity in OD of 20/60 and in OS of 20/200. With use of corrective lenses, he has achieved a BCVA in OD of 20/20 and in OS of 20/20. No corneal scars, corneal opacities, or Vogt's striae were shown on ocular biomicroscopy (slit-lamp examination). No significant data were shown on ocular motility, intraocular pressure, or funduscopy. The corneal topography and ultrasonic pachymetry data are shown in Table I. Keratometry in the right eye was 41.8 × 45.5 D, with greater curvature of 48.9 D, and in the left eye was 41.8 × 44.4, with a greater curvature of 46.5 D. Corneal thickness in OD was 507 microns and in OS was 511 microns.

A medical certificate was issued with validity restriction, requesting medical evaluation every 12 mo. No other operational limitation has been imposed.

## DISCUSSION

Keratoconus is a medical condition with aeromedical significance and should be reported to aviation medical examiners upon diagnosis. Certification in civil aviation is possible in cases where there is stable disease with a stable response to vision correction and treatment. These are evaluated based on the maintenance of BCVA and refractive parameters during the period of certification.

As the disease can potentially progress over a short period of time, certificate validity may be shortened accordingly, especially in affected younger applicants, whose disease could advance more aggressively. Alternatively, operational limitations can be imposed if the medical examiner allows a standard certification period. These measures mitigate the accident risk in airline pilots and commercial pilots operations.

Further studies might be considered in evaluating the risk of incapacitation in hypoxic and hypobaric conditions.<sup>2,3,12</sup> These studies should include evaluation of the risk of acute hydrops, which is significant when there are extremely thin corneas and significant corneal scarring.<sup>7</sup> It is a painful, disabling condition incurring the danger of sudden incapacitation, even if occurring in only one eye. In severe cases, even the use of glasses or rigid contact lenses will not be able to correct the visual acuity to what is acceptable for safe flying.

Distracting distortions, glare, flare, monocular diplopia, ghost images, unstable refraction, and uncorrectable refractive error are symptoms that deserve special attention and put keratoconus as a cause for incapacitation during flight. In these cases, where the pilots do not achieve a BCVA better than 20/30 in each eye and 20/20 with both eyes (airline pilots and commercial pilots), or present some of the symptoms above, they must not return to flying. None of the cases presented in this report had these predisposing conditions.

These criteria are also used after any form of surgical treatment for keratoconus.<sup>4,5</sup> As recovery time after surgical procedures varies individually, the period of returning to flying is analyzed on a case-by-case basis by Brazilian medical assessors of the civil aviation authority, with consideration of evaluation of BCVA and lack of symptoms. According to the civil aviation authorities mentioned above, even in cases where the applicant can meet the vision requirements for aeronautical activity, regular specialist review will be required.

International civil aviation authorities regulate the frequency of aeromedical and ophthalmic review of airline pilots and commercial pilots by no more than 1 yr (it can be shortened to 6 mo in some specific operational cases). As keratoconus, classically, has its onset at puberty and is progressive until the third to fourth decade, when it usually arrests, Brazilian medical assessors also consider pilot age, BCVA stability, and operational pilot category to maintain, or to shorten, the validity of medical aeronautical certification (from 6 mo to 1 yr).

The literature addressing sudden incapacitation due to this specific condition is lacking. Furthermore, these cases support the need for versatility and a broad knowledge base in the practice of aviation medical examiners to evaluate ocular pathology.

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