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13. ABSTRACT

Despite the rigid screening of all categories of rated personnel for the ability to meet stringent visual standards, cases of keratoconus continue to occur in the USAF flying population. This paper reviews the recent experience of the USAFSAM Aeromedical Consultation Service with this disease entity. Aspects of incidence, early diagnosis, serial progression, and treatment are discussed. Two cases of keratoconus which developed after four years of corneal contact lens wear are presented; and the increasing problem of intentional corneal molding ("orthokeratology") is reviewed.

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Keratoconus in USAF Flying Personnel

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Despite the rigid screening of all categories of rated personnel for the ability to meet stringent visual standards, cases of keratoconu continue to occur in the USAF flying population. This paper reviews the recent experience of the USAFSAM Aeromedical Consultation Service with this disease entity. Aspects of incidence, early diagnosis, serial progression and treatment are discussed. Two cases of keratoconus which developed after four years of corneal contact lens wear are presented; and the increasing problem of intentional corneal molding (orthokeratology) is reviewed.

THE UNITED STATES AIR FORCE has a continuing interest in the rehabilitation of all flying personnel who are found to have developed ophthalmologic disease. One such disease is keratoconus. Prior to the development of therapeutic techniques utilizing contact lenses, this condition was cause for unconditional grounding. Although the USAF visual standards for rated personnel in all categories are stringent, clinical cases of keratoconus continue to be seen despite rigid screening. If these clinical cases can be rehabilitated to continue in flight status after initial diagnosis, a significant savings accrues to the government, both operationally and economically.

This review examines the USAF School of Aerospace Medicine's recent experience with keratoconus in its rated aircrew personnel. Information was sought regarding factors of etiology which may have occurred commonly enough to be of predictive value, or those which might indicate the efficacy of treatment in selected cases, when adequately diagnosed.

Keratoconus is a condition characterized by a noninflammatory conic or cone-like protrusion of the cornea. It usually becomes manifest in adolescence and causes marked visual impairment secondary to the high degree of irregular myopic astigmatism which develops. It is difficult to cite the incidence of keratoconus in the general population but according to recent statistics from five large series, the median frequency of occurrence in eye patients is 1 in 413 (303/132,402 = 2.3%).[6] It is probable that the frequency would be even higher if the early abortive forms were diagnosed more frequently. In Amsler's[1,2] reports on 600 cases fully 52% were of the attenuated or rudimentary forms (formes frustes). These early forms often are not diagnosed, as visual acuity is generally normal. The disease is usually bilateral, manifesting itself initially about the time of puberty. A predilection for females noted in earlier series has not been a constant finding of recent authors, nor has the contention that the disease is rare in Negroes.[6,7] When it involves one eye it is usually slight in degree, and in most cases an asymmetry of the astigmatism betrays the presence of a minimal corneal deformity in the other eye also.[4]

The clinical course may be extremely variable. At the time of initial diagnosis it may be stationary, or it may progress either slowly or rapidly. Amsler[2] re-examined 366 eyes with keratoconus between three and eight years later. Only 87 (22.5%) had shown progression, and fully 33% of the early forms had been stationary.

The essential pathological lesion of keratoconus appears to be an abnormal yielding of the central portion of the cornea. The degree of thinning varies with the severity of the condition. Bowman's membrane is attenuated and there are gaps in its continuity. These gaps are filled with a type of connective tissue differing from the normal corneal stroma and containing elastic fibers.[3,5] It has been shown that the basal cells of the epithelium are also affected early, while other layers of the epithelial cells remain normal for a long period. Consequently ruptures in Bowman's membrane and degeneration of the collagen fibers appear, followed by connective tissue formation from the parenchyma.[8] Teng recently confirmed by electron microscopy that it is the basal epithelium which is the primary seat of the degenerative process. The liberation of proteolytic or autolytic enzymes then deleteriously alters the remainder of the cornea.[3,5] The subsequent loss of collagen

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may be the principal etiology for the cone-shaped appearance. Participation of the sclera in this process has been suggested, but its role may be just that of another expression of the defective development of the mesenchyme.

The role of heredity in the etiology of keratoconus remains an enigma. Its importance, however, is increasingly admitted. In the cases in which consanguinity has been noted, recessive inheritance appears likely. On the other hand there are many cases in which dominant and, more frequently, irregular transmission through two and even three generations is not uncommon.[5,11] The problem probably rests with the fact that the attenuated early forms (formes frustes) are so seldom recognized. This, coupled with the weak penetrance of the keratoconus gene and its highly variable expression, often leaves genealogical tracing in doubt. This explains why keratoconus of different degrees may appear not only in the same patient, but also in the same family, and why some carriers may show no clinical symptoms. Additional proof of the heredity factor is the concordance seen in monozygotic twins. Finally, the frequent association noted with other hereditary conditions makes this mode particularly suspect (e.g., mongolism, allergy, retinitis pigmentosa, infantile tapetoretinal degeneration, blue sclerae, etc.).[6]

TABLE I. USAF VISUAL STANDARDS*

Maximum Refractive Error

	Hyperopia	Myopia	Astigmatism
Flying Class I	1.75 Diopters In Any One Meridian	0.25 D In Any One Meridian	0.75 D In Any One Meridian
Flying Class IA	3.0 D In Any One Meridian	1.50 D In Any One Meridian	2.00 D In Any One Meridian
Flying Class II	3.50 D In Any One Meridian	3.50 D In Any One Meridian	No Type Over 3.00 D
Flying Class III	5.50 D In Any One Meridian	5.50 D In Any One Meridian	No Type Over 3.00 D

Minimum Visual Acuity

	Near		Distance	
	Uncorrected	Corrected	Uncorrected	Corrected
Flying Class I	20/20 Each Eye	———	20/20 Each Eye	———
Flying Class IA	20/20 Each Eye	———	20/30 Each Eye	20/20 Each Eye
Flying Class II	20/200 Each Eye	20/20 EachEye	20/200	20/20 Each Eye
		20/30 In	20/400	20/30 In
Flying Class III	No Standards	One Eye; 20/30 In The Other		One Eye; 20/30 In The Other

*AFM 160-1

TABLE II

Patient	Aeronautical Rating	Best Visual Exam. Category Passed
D.C.	Pilot	I
G.A.	Nav.	IA
J.C.	Other*	III
K.H.	Pilot/Nav.	I**
D.J.	Other*	III
D.M.	Nav.	IA
G.N.	Pilot	I**
J.S.	Pilot	IA**
S.W.	Nav.	IA
J.T.	Nav.	IA
T.W.	Pilot	I

* Enlisted aircrew specialties

** Includes two cases who cheated on flying class I exams after corneal molding with contact lenses, and one in which pilot training entry was waived to flying class IA standards.

MATERIAL AND METHODS

Eleven medical records of keratoconus patients seen between 1965 and 1970 were reviewed at the USAF School of Aerospace Medicine's Aeromedical Consultation Service. All but one of these cases were initially referred for evaluation of decreased visual acuity, which could no longer be corrected to 20/20 using either a phoropter or a trial lens case. Items of interest were the patient's medical and visual history, including his ability to pass visual screening tests upon entry to flight training, during this student training phase and serially throughout his operational flying career. Also obtained were data on family history, past history of physical examinations and an in-depth visual examination. This latter included near and distant visual acuity, with and without ocular correction if required, motility, manifest and cycloplegic refraction, slit lamp examination, direct ophthalmoscopy, Placido disc or keratoscopic evaluation and keratometry.

For the purposes of this study the diagnosis of early keratoconus was made by the USAFSAM Ophthalmology Service, using biomicroscopy, and was based on any combination of three of the following criteria:

1. A definite irregular distortion of the mires on examination with either a keratoscope, Placido disc, or the keratometer.
2. A thinning of the cornea at the apex of the cone.
3. Vertical lines (Vogt) seen in the deeper layers of the corneal stroma (presumably caused by a stretching phenomenon).
4. An increased visibility of the corneal nerve fibers, which form a network of grey lines interspersed with small dots.
5. Fleischer's ring, a line running around the base of the cone which may be pigmented deep yellow or green from the deposition of hemosiderin.

TABLE III

Patient	History of Allergy
D.C.	None
G.A.	Hay Fever
J.C.	None*
K.H.	None
D.J.	Hay Fever
D.M.	None
G.N.	None*
J.S.	Multiple food allergies; Severe reaction to horse serum (Tetanus Antitoxin)
S.W.	Seasonal Pruritic Rash; Intermittent Therapy over a 10 year period.
J.T.	None*
T.W.	None

* These cases did, however, relate phenomena such as a history of Rheumatic Fever, moderate Acne, and recurrent Furunculosis in childhood.

DISCUSSION

The only recent information on the incidence of keratoconus is that from clinical ophthalmological referral centers. These data, previously cited, suggest a median frequency of occurrence in eye patients of 2.3%.[6] Of the 3,050 patients seen by the Ophthalmology Service at the USAF School of Aerospace Medicine from 1965 through 1970, only 11 cases of keratoconus have been recorded (11/3050 = 0.36%). It is assumed, however, that these 11 cases represent the experience from the entire USAF flying population. This suggests that the USAF visual screening standards for the flying population do, in fact, identify the vast majority of those individuals with early irregular astigmatism which may progress to keratoconus (Table I).

This contention gains additional support when one regards the distribution of the clinical disease among the rated specialties, and when one considers the best visual category the affected individuals had previously attained (Table II). Of the 11 cases, five are rated pilots. In two of these cases, however, it was discovered that corneal molding had been undertaken using contact lenses, so that the Flying Class I visual examination could be passed. In fact, one patient gave a history of taking the initial pilot training physical and all subsequent interval physical examinations with his contact lenses on! In another case the Flying Class I minimums had been administratively waived to Class IA to allow an Air Force Academy cadet to enter pilot training. It is interesting to note that the remaining two cases both involve only one eye; one patient has evidence of old ocular trauma to the affected eye and the other is presently in remission.

Previous authors have increasingly admitted the role of heredity in the etiology of keratoconus, despite the fact that its exact mode of transmission has remained an enigma. Our 11 cases revealed no overt evidence of prior family involvement, but four cases had distinctly positive histories for allergic phenomena (4/11 = 36%). In addition three patients gave a history of having had rheumatic fever, moderate acne or recurrent furunculosis in childhood. While the allergic component of these conditions may be in doubt, their relationship here is interesting (Table III).

At the time of first referral to the USAF School of Aerospace Medicine, four of the 11 cases involved both eyes. Four additional cases were uniocular only and have remained so. (Although this group contains three cases with relatively short follow-up since initial diagnosis, and one case with evidence of earlier uniocular trauma). Air Force medical surveillance of these eleven cases ranges from 2 to 20 years, with an average of 9.5. Follow-up between the time of first diagnosis of keratoconus and 1970 (or until lost to follow-up via discharge, etc.) ranges from 11 to 66 months, with an average of 28.5. In the remaining three cases who developed keratoconus in the second eye while being followed at the USAF School of Aerospace Medicine, this involvement occurred between 11 and 31 months, following initial diagnosis, and averaged 19.7 months. Authors who have reviewed the onset of keratoconus in the general populace usually cite the time of onset as late puberty or early adulthood.[7] In our 11 patients the age at time of first diagnosis ranged from 21 to 38, with a median and average age of 28 years.

The basis for diagnosis in this study was adapted from classic authors to aim at the earliest possible identification. The advanced stages of this disease were not expected in our population, and indeed, in only one case was stigmata of severe disease found (Table IV). This may include ruptures in Descemet's or Bowman's membranes, with or without the development of "acute keratoconus." This latter entity is not an acute development of kertoconus, but rather an acute pseudo-inflammatory exacerbation in subjects who have keratoconus.[15] "Acute keratoconus" is seen when the cornea is infiltrated by aqueous humor after a rupture of Descement's membrane and the endothelium. A sudden deterioration of vision, marked corneal edema and greatly aggravated bulging of the corneal stroma are hallmarks of this complication.[3] Only rarely does the ectasia involve the whole of the cornea, giving rise to a condition resembling keratoglobus.

In every case the problem which caused referral to USAFSAM was decreased visual acuity secondary to the corneal changes. In only one case could 20/20 visual acuity be restored with glasses. This one case involved an RF-4C pilot who complained of blurred vision, but who could achieve 20/20 vision by head tilting. The best phoropter-corrected near and distant visual acuity in the affected eyes ranged from 20/25 to 20/40, with a median of 20/30.

After detailed evaluation of these 11 cases it was determined that each had been diagnosed early enough by USAF medical surveillance to warrant a trial fitting with contact lenses. In all but one case 20/20 visual acuity was attained after careful corneal contact lens fittings. However, two patients were unable to adapt well enough to the wearing of contact lenses to justify their continued use; both of these patients were permanently grounded. In the remaining nine cases the lenses were well tolerated and the recommendation was for return to flying duties. This group included the one navigator who could not be corrected better than 20/25 in one eye. All of these nine cases resumed unrestricted aircrew duties without difficulty. The follow-up period on this latter group varies from 12 to 66 months and averages 26.7. Two of these have been discharged or retired; the remaining seven are presently performing active flying duties, with three wearing only a single contact lens.

Although problems associated with the use of con-

TABLE IV

Factor	Number Having The Factor	Percent
1. Marked distortion of the Mires	11	100
2. Thinning of the cornea	8	73
3. Increased prominence of the corneal nerves	7	64
4. Vertical stress lines	7	64
5. Fleisher's Rings	2	18
6. Rupture in Descemet's Membrane	1	9
7. Rupture in Bowman's Membrane	0	0

rective corneal contact lenses have long been known,[8,10,12] including the spectre of the lenses themselves actually giving rise to keratoconus,[9] it is probably prudent to suppose that this mode of contemporary therapy is the most efficacious for this particular disease.[7,13] The size of this series limits any discussion regarding complications seen with corneal contact lenses; but it is interesting to note that in two of our cases of documented keratoconus, a history of wearing corneal contact lenses for four years was discovered. As with Hartstein's initial four cases with this complication, first reported in 1968 in the Archives of Ophthalmology,[9] it is impossible to state whether the keratoconus would have occurred by itself, or if it was actually induced in some manner by the wearing of the corneal contact lenses.

CONCLUSIONS

The development of keratoconus in USAF flying personnel remains a persistent problem. This study of cases referred over a six-year period to the USAF School of Aerospace Medicine suggests, however, that the present USAF visual standards are adequate to screen out the majority of potential keratoconus patients, particularly in the pilot population. The need to take a careful visual history is once again emphasized. The technique of intentional corneal molding with contact lenses, generally called "orthokeratology," will undoubtedly cause the flight surgeon increasing problems in the future. An awareness of the technique and its increasing popularity should alert the examining physician to search for it in all flying applicants. If corneal molding is suspected, serial refractions and keratometry after contact lens removal should be accomplished. The necessity for a detailed ophthalmological examination of all flight training applicants, and a particularly careful review of past refractive data whenever available, seems imperative. The advisability of granting waivers for decreased visual acuity, particularly if the parameter in question is irregular astigmatism, is questionable. By use of the present USAF visual standards, precisely applied in every case, the best use of available resources can be expected.

If keratoconus does develop, and if early diagnosis is made, the majority of aircrew personnel can be returned to operational flight status using corrective corneal contact lenses. The importance of serial follow-ups to avoid complications, at least every three months initially, is stressed. Finally, this salvage of rated personnel from what had long been held to represent a mandatory grounding condition represents a significant savings both to the individual flyer and to the United States Air Force.

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