# Life expectancy in keratoconus

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### **Abstract**

It is observed that few patients over the age of 60 regularly attend the keratoconus clinic at Moorfields Eye Hospital. The hypothesis that patients with keratoconus have a shorter life expectancy owing to underlying connective tissue related disease was tested. From patient records a sample of 337 keratoconus patients aged at least 40 years by 1991 were identified, of which 279 were living, 13 were deceased, and 45 were untraceable. The mortality rate for keratoconus patients was compared with that of the general population using actuarial English life tables. Results show no significant difference between the general population mortality rate and that of the keratoconus sample even with adjustment for social class. Possible explanations for the non-attendance of older patients are discussed.

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The keratoconus clinic at Moorfields Eye Hospital now has more than 2000 registered patients and it is observed that most patients seen are aged under 40 years and that very few patients over 60 years are seen regularly.

There are several reports linking keratoconus and connective tissue disorders such as Ehlers-Danlos syndrome, osteogenesis imperfecta, and mitral valve prolapse. Could it be that patients with keratoconus have a shorter life expectancy owing to underlying connective tissue related disease or is there an alternative explanation? The purpose of the present study was to investigate this matter.

## Patient sample and method

The keratoconus clinic at Moorfields Eye Hospital was established in 1972 and from patient records we were able to identify a sample of 862 new patients with keratoconus who presented between June 1972 and December 1982. Of these 71 had no recorded date of birth and were excluded from the sample. From the remaining 791 those born prior to 1951 were identified as they would now be at least 40 years of age. This sample consisted of 337 patients and, of these, 112 were still attending the clinic leaving 225 unaccounted for. We attempted to trace these patients in order to determine why they were no longer attending the clinic. Those that we were unable to contact using patient records were identified through the Office of Population Censuses and Surveys as being dead or alive. Where possible the cause of death was determined. By these methods we were able to obtain conclusive data on 178 of the 225 patients no longer attending the clinic. The remainder (47) were classified as untraceable and included those patients who were known to have moved abroad. However had they died in the United Kingdom they would have been traceable.

#### Results

Of the study sample of 337 keratoconus patients aged at least 40 years by 1991, 279 were living, 13 were deceased, and 45 were untraceable. The sample comprised 184 males (mean age 57.75 years, range 41–79) and 153 females (mean age 51.83 years, range 41–83). The ratio of males to females was 1.20:1.00; 23.4% of the sample were aged over 50 years.

In order to assess whether the mortality for keratoconus patients was greater than that of the general population actuarial English life tables were used. These tables are used to assess general population mortality rates and the table used (ECT14) is based on the mortality experience of England and Wales during the period 1980–82 – that is, the year in which the sample was complete. From it we were able to calculate the predicted general population mortality rate by 1991 and compare it with that of the keratoconus sample. Male and females were assessed separately as their mortality rates differ (Tables 1 and 2).

There was no significant difference between the general population mortality rate and that of the keratoconus sample [ $\chi^2=1.59$  (males),  $\chi^2=1.31$  (females)].

It is known that male mortality in Great Britain differs according to social class.<sup>3</sup> It has been shown that the social class distribution of patients attending the keratoconus clinic at Moorfields Eye Hospital is not typical of the general population.<sup>4</sup> In view of these findings the figures were recalculated for social classes I and II, as these groups constituted 77% of the patients attending the clinic. The figures for the

Table 1 Females

Year of birth	Number 1982	Predicted 1991	Actual 1991
50	12	11.9	11
49	12	11.89	12
48	6	5.94	5
47	7	6.92	7
46	5	4.94	5
45	2	1.97	2
44	6 75 2 8 5 7 3 2 6 5 3 2 2 4	7.88	5 7 5 2 8 5 7 3 2 6 5 3 2 2
43	5	4.91	5
42	7	6.87	7
41	3	2.93	3
40	2	1.95	2
39/8	6	5.84	6
37	5	4.84	5
36	3	2.89	3
35/4	2	1.92	2
33	2	2.86	2
1903/1913 (1908)	. 4	2·31	1
1914/1932 (1923)	9	7.98	9
Total	110	96·74	104

χ<sup>2</sup>=1·31; not significant

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Table 2 Males

Year of birth	Number 1982	Predicted 1991	Actual 1991	
50	21	20.7	21	
49	30	29.6	30	
48	20	19.72	20	
47	16	15.75	16	
46	17	16.66	17	
45	10	9.8	10	
44	9	8.80	9	
43	6	5.85	6	
42	12	11.66	12	
41	4	3.86	4	
40	3	2.89	2	
39	5	4.8	5	
38	3	2.88	2	
37	3	2.88	3	
36	5	4.74	5	
35	4	3.77	4	
34	12 4 3 5 3 3 5 4 2 3	1.85	2	
33	3	2.76	2	
1932/1920 (1926)	15	12.6	9 6 12 4 2 5 2 3 5 4 2 2 2 13	
1913/1910	2	1	2	
Total	190	181.89	185	

 $\chi^2 = 1.59$ ; not significant

Table 3 Males (adjusted for social class after Haberman and

Year of birth	Number 1982	Predicted 1991	Actual 1991	
50	21	20.81	21	
49	30	29.71	30	
48	20	19.79	20	
47	16	15.82	16	
46	17	16.77	17	
45	10	9.85	10	
44	9	8.86	9 6 · 12	
43	6	5·89 11·76		
42	12			
41	4	3.91		
40	4 3 5 3 3 5 4 2 3 15	2.92	2	
39	5	4.85	5	
38	3	2.90	2	
37	3	2.88	3	
36	5	4·78 3·80	4 2 5 2 3 5 4 2 2	
35	4			
34	2	1.89	2	
33	3	2.81	2	
1932/1920 (1926)	15	12.96	13	
1913/1910 (1912)	2	1.04	2	
Total	190	184	185	

 $\gamma^2 = 0.53$ ; not significant

Table 4 Males

Year of birth	Age at death	Cause
1949	36	Accident (flying)
1938	47	Status asthmaticus
1934	46	Asphyxia; food in larynx (mentally retarded)
1940	45	Respiratory failure. Pulmonary sarcoid
1925	63	Ca larynx with 2°
1920	66	Ca bladder
1933	57	Ca oesophagus
Females		
1905	78	Carcinomatosis
1903	70	Pulmonary embolism following RTA
1950	24	Suicide: acute barbiturate poisoning
1907	82	Ca bronchus: bronchopneumonia
1948		)
1933	; ;	Myocardial infarction

Ca=cancer.

social class adjusted males are given in Table 3. Again, no significant difference was found between the general population mortality rate and that of the keratoconus sample ( $\chi^2 = 0.53$ ). Figures for female mortality based on social class are not available.

Data on the deceased patients obtained from death certification or the patients' general practitioners are summarised in Table 4.

## Discussion

Our results show that both male and female keratoconus patients do not have a shorter life expectancy than the general population. With regard to the male patients, even after the data were adjusted to allow for social class differences, there was still no significant difference. Thus our hypothesis that underlying connective tissue disorder shortens the life of keratoconic patients is not supported. However it must be remembered that patients with multi-system disease such as Down's syndrome may not be referred to Moorfields in view of their low success rate in contact lens wear.

The observation that most of the patients attending our keratoconus clinic are aged 40 years or less may be explained by other factors, including:

- (1) older patients no longer attend because they are satisfied with their spectacle or contact lens correct vision, and do not consider that they require medical surveillance of their ocular condition;
- (2) it is known that for a given patient the natural history of the disease is independent between the two eyes.5 Thus the disease may have progressed very little if at all in the better eye and the older patients may be content with, or are resigned to, somewhat subnormal vision and no longer wear their spectacles or contact lenses;
- (3) older patients who live out of London are seen by ophthalmologists or optometrists near their homes, being no longer willing to travel to London.

We plan to extend this study into future years as, though we have detected no indication of reduced life expectancy, the sample remains fairly young, less than a quarter of the patients being aged over 50 years.

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<sup>1</sup> Maumenee IH. Hereditary connective tissue disorders involving

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<sup>115: 495</sup> 

<sup>4</sup> Woodward EG. Keratoconus: maternal age and social class. Br J Ophthalmol 1981; 65: 104-7.
5 Woodward EG, Moodaley LC, O'Hagan A. Predictors for likelihood of corneal transplanation in keratoconus. Eye 1990; 4: 493-6.