

## Hoarse voice resulting from premature ageing in Werner's syndrome

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### Abstract

Werner's syndrome is characterized by clinical signs of premature ageing. A 42-year-old man presented with three-year history of hoarseness. Also noted were skin atrophy of the face and hands, ulcerations around the ankles, and a history of cataracts. A clinical diagnosis of Werner's syndrome was made. Laryngoscopy revealed bowed vocal folds resulting in a spindle-shaped defect with glottal incompetence during phonation. Examination also revealed decreased maximum phonation time and vocal fatigue. At surgery, atrophy of the vocalis muscle was noted. Furthermore, degeneration of muscle fibres was noted in the temporalis muscle. The atrophic changes in the vocal folds that occur with ageing and result in an increased fundamental frequency were seen in this patient. The characteristic hoarseness of Werner's syndrome appears to be the result of premature ageing of the vocal folds.

**Key words:** Werners syndrome; Dysphonia

### Introduction

Werner's syndrome (WS) is an inherited disorder which is characterized by clinical signs of premature ageing including hair loss, cataracts, atrophy of the skin and peripheral fat, and diabetes. In addition, there are unusual clinical features unrelated to ageing, including ulcerations around the ankles and soft tissue calcification.

The incidence of this disease is highest in Japan. Of the 1100 patients reported world-wide, 810 have been Japanese.<sup>1</sup> Recently, the gene responsible for Werner's syndrome was identified by positional cloning,<sup>2</sup> well after the disorder was named and the pathological changes were known. However, the syndrome is not completely understood. In this paper, a patient is presented in whom the initial manifestation of Werner's syndrome was hoarseness. The similarities in the vocal fold changes in Werner's syndrome and ageing are described.

### Case report

A 42-year-old man was referred to our clinic by his family physician because of a three-year history of progressive breathy hoarseness which resulted in a high-pitched hoarse voice. Laryngoscopy revealed bowed vocal folds resulting in a spindle-shaped defect with glottal incompetence during phonation (Figure 1). The anatomical and functional problems which resulted in incomplete glottal closure during phonation led to the breathy hoarseness, decreased maximum phonation time, and vocal fatigue.<sup>3</sup> In general, this type of vocal fold atrophy is the result of the ageing.

The patient had a history of bilateral cataracts which had been treated surgically two years earlier. In addition, there was a two-year history of ulceration around the

ankles (Figure 2). Skin atrophy of the face and hands was noted on examination (Figure 3). A clinical diagnosis of Werner's syndrome was made. Autologous transplantation of fascia into the atrophic vocal folds,<sup>4</sup> was performed to treat the voice disorder. The phonosurgical technique (Figure 4) comprised three steps.

- (1) undermining under the vocal fold mucosa to make a pocket inside the vocal fold;
- (2) preparation of fascia temporalis;
- (3) autologous (fascia temporalis) transplantation into the vocal fold.

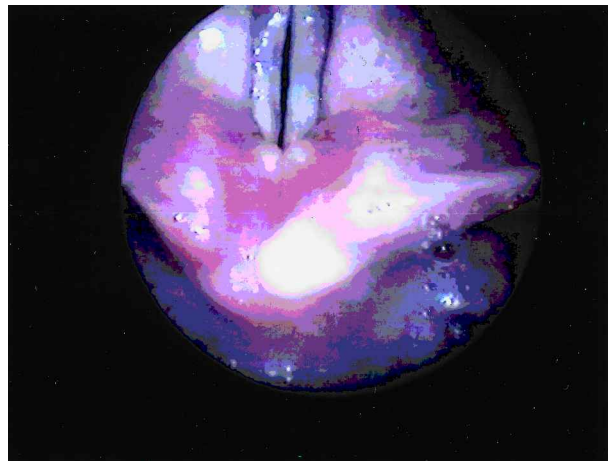


FIG. 1

Laryngoscopic view of the larynx. On phonation; note the glottal incompetence resulting from bowing of the vocal folds

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FIG. 2

Photograph showing the ulcerations on the atrophic ankles.

The breathy hoarseness and vocal fatigue resolved, and the maximum phonation time was increased from three to 15 seconds 12 months after the surgery.

### Discussion

A hoarse high-frequency voice is a well-known feature of Werner's syndrome. Surgical exploration in this patient demonstrated atrophy of the vocalis muscle. This contributed to the spindle-shaped vocal fold deformity and resulted in the husky voice. At the time of surgery, we

### Autologous Transplantation of Fascia into the Vocal Fold

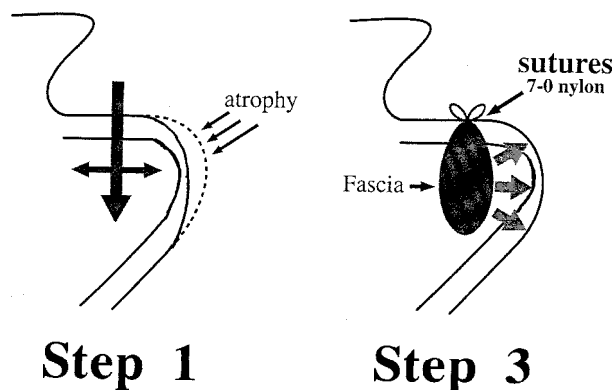


FIG. 4

Autologous transplantation of fascia into the vocal fold.



FIG. 3

Photograph showing the atrophic changes in the face characteristic vogelgeschicht.

removed a small piece of temporalis fascia with temporalis muscle. Degeneration of temporalis muscle fibres was recognized. These changes would normally be seen with ageing (Figure 5).

It is not uncommon for older men to exhibit marked vocal fold atrophy, which result in a higher fundamental frequency than is found in young men.<sup>5</sup> Voice changes in senescence are characterized by slight hoarseness and a noticeable change in fundamental frequency. A change in the mass of the vocal folds due to atrophy is considered to be responsible for these voice changes. Mysak<sup>6</sup> has compared middle-aged sons to their elderly fathers, noting

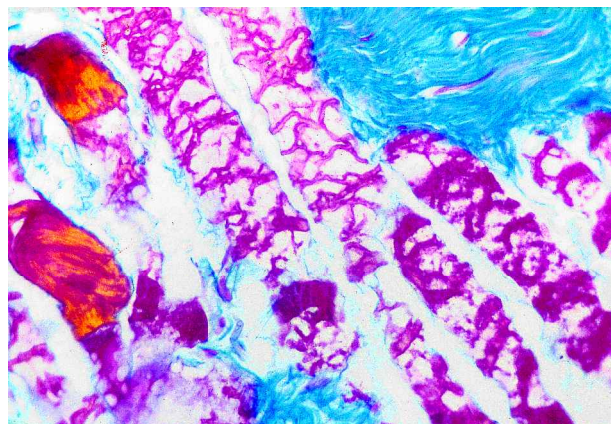


FIG. 5

Degeneration of muscle fibres in the temporalis muscle (Azan;  $\times 400$ ).

an increasing fundamental frequency of speech with advancing years starting in middle age. Hollien and Shipp<sup>7</sup> have studied men aged 20 to 80 and, in addition to confirming this trend, they established that the fundamental frequency curve was saucer-shaped when younger subjects were included. They reported that the vocal folds are thickest in the fifth and sixth decades, and that the increasing fundamental frequency is secondary to vocal fold thinning and stiffening. The finding in this case suggest that the characteristic hoarseness of Werner's syndrome is the result of premature ageing of the vocal folds.

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