

# Ultrasound Biomicroscopic Findings in Hallerman-Streiff Syndrome

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**Purpose:** To demonstrate the usefulness of ultrasound biomicroscopy in detecting the morphological changes in the lens caused by the spontaneous absorption of lens material and to detect fundus abnormalities in a patient with Hallermann-Streiff syndrome.

**Methods:** Case report of an infant diagnosed at the age of 2 months as having Hallermann-Streiff syndrome.

**Results:** Spontaneous lens absorption occurred during the course of follow-up and was detected only by ultrasound biomicroscopy after the patient was prepared for cataract surgery. The changes in the anterior chamber depth and lens shapes were documented by ultrasound biomicroscopy. Retinal folds that were barely observable by conventional ophthalmoscopy because of a dense cataract were clearly shown by ultrasound biomicroscopy.

**Conclusions:** Ultrasound biomicroscopy can be used to examine the lenses of eyes that are not observable with conventional optical instruments. Ultrasound biomicroscopy can also be used to study the posterior segment of microphthalmic eyes. We recommend preoperative ultrasound biomicroscopy to prevent unnecessary anesthesia and surgical preparation. **Jpn J Ophthalmol 2002;46:451-454** © 2002 Japanese Ophthalmological Society

**Key Words:** Congenital cataract, Hallermann-Streiff syndrome, microphthalmos, retinal fold, ultrasound biomicroscopy.

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## Introduction

The Hallermann-Streiff syndrome is a rare congenital anomaly first described by Hallermann<sup>1</sup> and Streiff.<sup>2</sup> The most common features of this syndrome are dyscephalia, dental anomalies, hypotrichosis, cutaneous atrophy, and dwarfism. Various ophthalmic findings have been reported, and the most common findings are congenital cataracts (81–90%) and microphthalmia (78–83%).<sup>3</sup> Spontaneous lens absorption sometimes occurs in older patients. At first, this process may lead to a temporary improvement in vision, but later in life, chronic, intractable iridocyclitis may develop and cause many complications. Histopathological studies have shown shrunken cataracts and spontaneous aphakia with or without iridocycli-

tis around the empty capsule.<sup>4,5</sup> Hopkins and Horan recommended early surgical removal of the lens, rather than waiting for a spontaneous lens absorption, to prevent secondary glaucoma.<sup>6</sup>

We report a case of Hallermann-Streiff syndrome in which spontaneous lens absorption was detected only by ultrasound biomicroscopy.

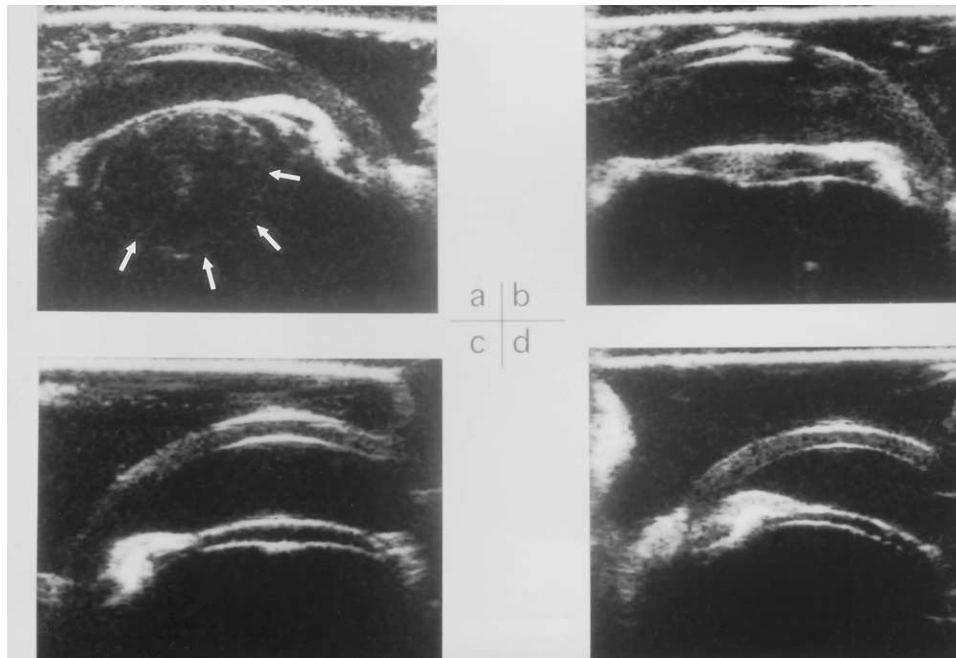
## Case Report

A 2-month-old Japanese boy was referred to the Nagoya University Hospital. He was born prematurely at a gestational age of 32 weeks and weighed 1,624 grams. Because of the characteristic bird-like face, he was diagnosed as having Hallermann-Streiff syndrome. On examination, the corneal diameter was 7 mm horizontally OU (9–10 mm is normal), and the corneas were clear in slit-lamp microscope examination. A posterior embryotoxon was present with a high insertion of the iris and a posterior synechia, OU. The anterior chamber was

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**Figure 1.** Ultrasound biomicroscopy findings (30 Hz) at 3 months of age (**a,b**) and at 5 months of age (**c,d**) in a patient with Hallermann-Streiff syndrome. (**a**) Right eye at 3 months; shallow anterior chamber, anomalous angle, and round-shaped lens can be seen (arrows). (**b**) Left eye at 3 months; shallow anterior chamber, anomalous angle, and membranous lens can be seen. (**c**) Right eye at 5 months; lens is thin and membranous. (**d**) Left eye at 5 months; lens is thinner than before.

very shallow in the right eye and moderately shallow in the left eye. This is a characteristic finding in microphthalmic eyes.

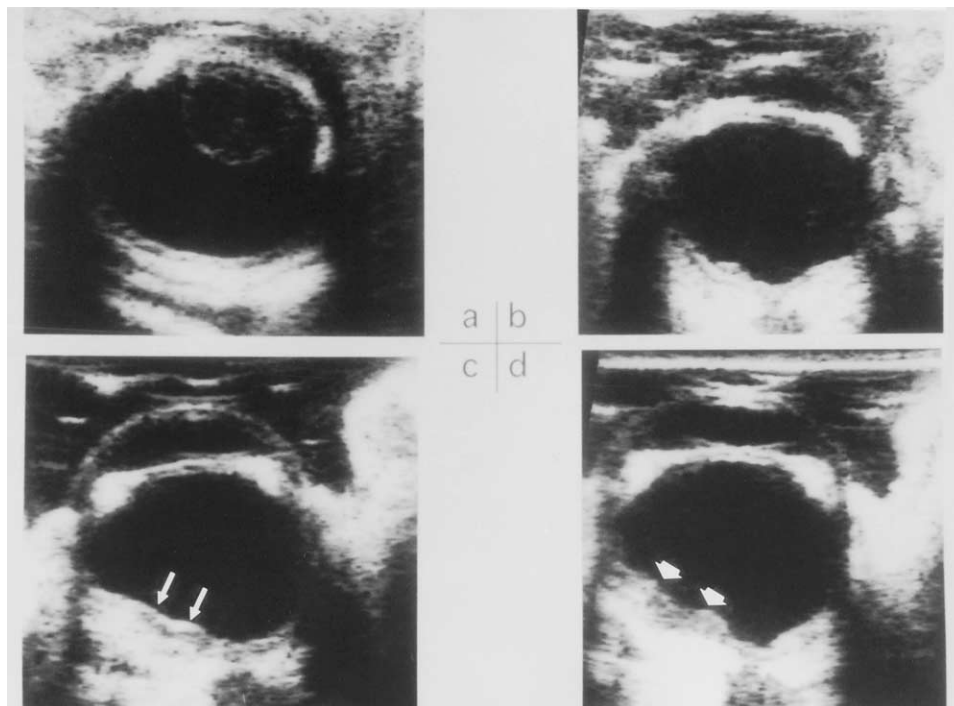
The right lens was cataractous, and the fundus was not visible because of poor dilation of the pupil. The left lens showed cataract punctata, and severe retinal folds were observed around the optic nerve head.

High-frequency ultrasound biomicroscopy (UX-2; Rion, Tokyo) was performed at two frequencies, 30 Hz for the anterior part of the eye (Figure 1) and 15 Hz for the whole eye (Figure 2). The axial length was approximately 9 mm, OU. The optic nerves were of normal size, and the sclera was 1-mm thick. The surface of the retina was smooth in the right eye but irregular in the left eye, a reflection of the retinal folds. The lens of the right eye was almost round and 3.2-mm thick and occupied about one third of the vitreous cavity. The anterior chamber angle was narrow and the distinction between cornea and sclera was obscure. The left lens was only about 0.8 mm thick and the anterior chamber angle was wider than in the right eye. There was no evidence of inflammation in the anterior chamber of either eye. No changes were visible for 2 months by slit-lamp microscopy. The patient did not

fixate on light, but he blinked with strong light; therefore, he was suspected of having light perception.

Surgery was planned at 5 months of age to remove the cataract in the right eye, as recommended by Hopkins and Horan to prevent secondary glaucoma.<sup>6</sup> Under general anesthesia, the intraocular pressure was 10 mm Hg OU. When ultrasound biomicroscopy was repeated before surgery, a thin image of the lens was observed in the right eye, suggesting that spontaneous absorption of the lens had occurred. This had not been detected by slit-lamp microscopy. The anterior chamber angle was wider than at the initial ultrasound microscopy examination. The right fundus was still not visible because of the poor dilation of the pupil and the opacity of the lens capsule. Moreover, the retina was not seen as smooth by ultrasound biomicroscopy as it had been at the previous examination, suggesting the formation of retinal folds as in the left eye. Subretinal fluid was observed by fundus examination and by ultrasound biomicroscopy, and the fluid shifted when the patient's position was altered, suggesting a uveal effusion. The surgery was aborted because of the spontaneous absorption of the lens.

The patient suffered a respiratory arrest and died when he was 9 months old.



**Figure 2.** Ultrasound biomicroscopy findings (15 Hz) at 3 months of age (**a,b**) and at 5 months of age (**c,d**). (**a**) Right eye at 3 months. Microphthalmos with relatively large lens can be seen. (**b**) Left eye at 3 months. Microphthalmos and membranous lens with normal-sized optic nerve can be seen. (**c**) Left eye at 5 months. Thick retinal folds (arrows) are observed in the supine position. (**d**) Left eye at 5 months of age. Subretinal fluid has shifted toward the temporal side when body was tilted so that the temporal side is down.

## Discussion

The importance of ultrasound biomicroscopy in investigating the morphology of the anterior segment has been reported because this technology was developed 10 years ago.<sup>7</sup> Ultrasound biomicroscopy has also been used for examining the anterior chamber angle,<sup>8,9</sup> and sclerocornea in infants,<sup>10</sup> and its usefulness for pediatric eye examinations has been accepted. A spontaneous absorption of cataracts may be recognized by slit-lamp microscopy in some cases but, as in this case, the absorption may not be certain because of the dense opacity of the lens capsule and poor dilation of the pupil. However, the absorption of the lens was disclosed with certainty by ultrasound biomicroscopy, and even the fundus abnormalities that had not been noted earlier were detected. If the ultrasound biomicroscopy had been performed before anesthesia for the surgery, the patient would not have had to be anesthetized and prepared for cataract surgery.

It is noteworthy that the spontaneous absorption of the lens in Hallermann-Streiff syndrome was demonstrated by ultrasound biomicroscopy. This investigative technique also demonstrated its usefulness in

detecting fundus abnormalities in microphthalmic eyes. We recommend preoperative ultrasound biomicroscopy to prevent unnecessary anesthesia and surgical preparation.

## Conclusion

This study emphasizes the importance of ultrasound biomicroscopy as a noninvasive means of examining structures that are not visible by conventional optical instruments in infants.

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