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Case Report

# Superior canal dehiscence syndrome in children – A case report

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ABSTRACT

Initially described in 1998, superior semicircular canal dehiscence syndrome (SCDS) has become a well-studied neurootologic entity in adults by now. Unfortunately, experience with children is limited and a diagnostic and therapeutic algorithm is lacking. The article therefore wants to provide an overview of the existing literature on superior semicircular canal dehiscence syndrome in children. Furthermore a diagnostic algorithm for daily clinical life based on a case report from an eleven-year-old girl is presented.



## 1. Introduction

In 1998, Minor et al. described a new vestibular disease termed superior semicircular canal dehiscence syndrome (SCDS) for the first time [1]. In this disease, the solid osseous covering the superior semicircular canal of the vestibular labyrinth is absent. This dehiscence forms a "third" pathological window of the inner ear next to the oval and round windows that can cause a variety of auditory and vestibular symptoms [2,3]. These may include pronounced autophony, dysequilibrium, debilitating vertigo episodes, and even conductive hearing loss. While the disease is now well-researched and well-established in adults, and diagnostic and therapeutic procedures exist, there is limited knowledge of children diagnosed with SCDS. This paper provides an overview of the current literature on the superior canal dehiscence syndrome in children and demonstrates a possible diagnostic approach using a clinical case report.

## 2. Case report

### 2.1. Medical history

An 11-year-old girl was referred to the medical sleep center at the University ENT Clinic, Mannheim with suspected childhood obstructive sleep apnea. In addition to her sleeping medical history, she reported that she started to feel dizzy while playing the flute recently and her eyes began "to hop" as well. No hearing impairment, hearing loss, tinnitus, or autophony was specified. Furthermore, the young patient stated that she could not freely ride a bicycle yet. Past medical history neither indicates a head trauma nor antibiotic therapy with potentially ototoxic drugs.

### 2.2. Equipment-free diagnostics

In the exploratory ENT medical examination, the clinical head-impulse test according to Halmagyi and Curthoys [4] showed minimum catch-up saccades when tested in the right horizontal plane. Intact extraocular motor function was observed, no spontaneous or provoked nystagmus was detectable, and regular eye movements without saccades were observed. Hennebert's sign was positive: with the Valsalva maneuver, rotational eye movements could be evoked. During this maneuver, the girl felt dizzy. Tullio's phenomenon could not be triggered. The ear microscopy showed irritation-free ear canals and eardrums on both sides. All other ENT medical examinations were normal, considering the patient's age. She is scheduled for further neurootological diagnostic procedures in the interdisciplinary Dizziness Center after completion of the diagnostic and therapeutic interventions for childhood obstructive sleep apnea.

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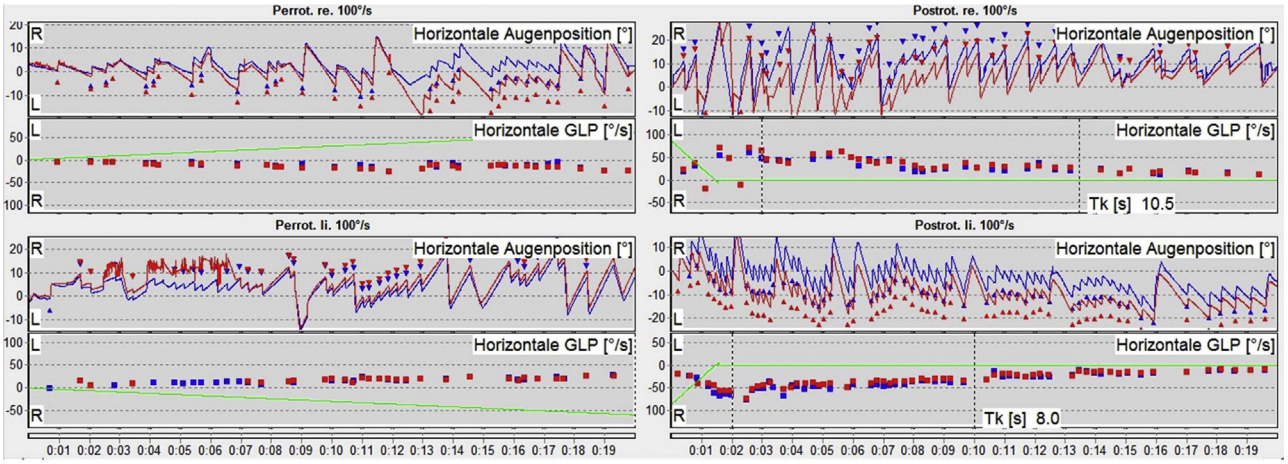


**Fig. 1.** Rotatory vestibular test: approximately the same bilateral excitability in pre- and post-rotatory testing.

### 2.3. Instrumental diagnostics

#### 2.3.1. Pure tone audiometry, rotary vestibular test, and caloric test

In the apparative diagnostics, an age-appropriate hearing ability was observed in pure tone audiometry. A differentiated receptor function analysis of the vestibular system was carried out.

The horizontal vestibuloocular reflex (VOR) was examined in the low-frequency range up to 0.19 Hz by means of a rotary vestibular and caloric test. The first rotary vestibular examination was conducted in an optimum position with binocular video nystagmography using the VNG VO425 FireWire™ system developed by Interacoustics A/S Denmark™ with a frame rate of 105 Hz [5]. The horizontal semicircular canal is usually provoked first by a weak stimulus and then a strong stimulus. The weak stimulus given initially consists of an acceleration of $2^\circ/s^2$ for 100 s up to a final speed of 200°/s. After a phase with a constant speed of 200°/s, a strong stimulus follows with a deceleration at the rate of $60^\circ/s^2$ to zero within 3½ s. The angular acceleration spectrum is therefore between $2^\circ/s^2$ and $-60^\circ/s^2$. Both in pre-rotatory and post-rotatory tests, no evidence of an asymmetric excitation or prolonged or disturbed post-rotatory recovery period was seen (Fig. 1). In the caloric test, measurement was performed with the same binocular video nystagmography device. The vestibular organ was triggered separately on both sides with warm water at 44 °C and cold water at 30 °C. In the automated nystagmus analysis, a side difference of 19% to the left and a directional preponderance of 22% to the left were seen (Fig. 2).

#### 2.3.2. Video head impulse test

Horizontal VOR was measured using a portable lightweight VOG device (EyeSee Cam™ system Interacoustics A/S Denmark™) that consists of a high-speed infrared camera (sampling rate of 250 Hz) and a built-in accelerometer. vHIT was performed with the girl sitting approximately 1.5 m away from a target mounted on the wall. Calibration was performed with integrated laser dots projected onto the wall.

The target head velocity was 100–200 degrees per second with an amplitude between 5 and 15° from the center to the lateral. The impulses were unpredictable with respect to both directions (right versus left). Twenty-three impulses to the right and 21 impulses to the left were performed. VOR gain was calculated as the ratio of the eye-to-head velocity at 40, 60, and 80 ms. Furthermore, the

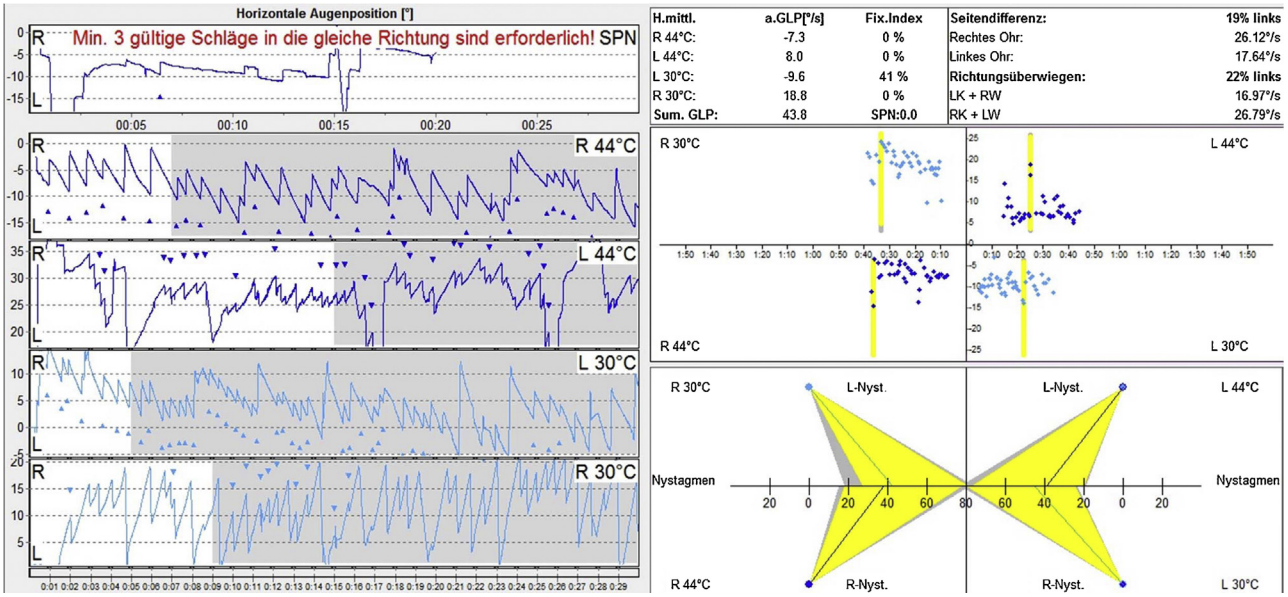


**Fig. 2.** Results of caloric testing: on the right side, the results of the test with hot and cold water are summarized separately for each side. On the left side, the evoked nystagmuses are given as an example.