

AUDIOLOGICAL SURVEILLANCE FOR USHER SYNDROME

PROVIDED BY: MONASH HEALTH LIBRARY**DATE:** 23 FEBRUARY 2026

Please find following a summary of a literature search and relevant results. All articles can be provided in full - email library@monashhealth.org for a list of the articles you require.

QUESTION

What is the current best practice for audiological surveillance of patients with Usher syndrome?

RESULTS

ONLINE RESOURCES

SCREENING GUIDELINES

Department of Health, Disability, and Ageing. (2025). **National Framework for Newborn Hearing Screening.** [Link.](#)

- Describes a 'screening pathway, including potential outcomes after a baby is referred for audiological assessment and discharged from the screening program' pg.5, fig.2
- Children with Usher Syndrome are considered eligible for screening pg.7

Ministry of Children, Community, and Social Services, Ontario, Canada. (2019). **Protocol for audiological surveillance of children at risk for permanent hearing loss.** [Link.](#)

- Lists Usher Syndrome as a group 2 risk indicator
- Group 2 risk indicator category recommends AABR screening, passes are recommended basic surveillance and an audiology visit at 15–18 months, failures are referred immediately to full audiological assessment pg.11-12

Coalition of Ohio Audiologists and Children's Hospitals. (2016). **Recommended Protocols for Diagnostic Audiological Assessment Follow-up to Newborn Hearing Screening in Ohio.** [Link.](#)

- 'Standardized diagnostic evaluation measures for infants who did not pass their newborn hearing screening' pg.1
- 'Earlier and more frequent monitoring is recommended (eg. every 6 months)' for children with Usher syndrome or other 'high-risk factors' pg.15

Health and Social Policy, NSW Gov. (2010). **Statewide Infant Screening - Hearing (SWISH) Program** [Link.](#)

- Usher Syndrome associated with progressive or late-onset hearing loss, therefore eligible for SWISH AABR screening and require ongoing audiological monitoring pg.60
- SWISH AABR screening protocol and referral flowchart pg.19

EDUCATIONAL MATERIAL

Cleveland Clinic. (2024). **Usher Syndrome**. [Link](#)

- Diagnosis: ‘Hearing tests: An ear, nose and throat specialist (otolaryngologist) and an audiologist will measure your child’s hearing’
- Management: Cochlear, Hearing aids, Vision aids, Early intervention

Better Health Channel, Vic Gov. (2019). **Usher Syndrome**. [Link](#).

- Broad Usher syndrome diagnosis and management overview

National Institute on Deafness and Other Communication Disorders. (2017). **Usher Syndrome**. [Link](#).

- Emphasises the importance of early diagnosis pg.3
- ‘Treatment and communication services may include hearing aids, assistive listening devices, cochlear implants, auditory (hearing) training, and/or learning American Sign Language.’ pg.3

Usher Kids Australia. **Ear**. [Link](#).

- Proposed management pathway: Progressive audiological assessment, Early fitting of hearing aids/Cochlear implant, AUSLAN

PEER-REVIEWED JOURNAL ARTICLES – MOST RECENT FIRST

Each article summary contains excerpts from the abstract and an online link.

Başöz Behmen, M., et al. (2026). **Audiological findings in usher syndrome according to types**. *The Egyptian Journal of Otolaryngology*, 42, 2. [Full text](#)

The aim of this review is to compare the auditory findings observed according to different types of Usher syndrome in the literature and to reveal effective treatment and follow-up management. It also concludes that that genetic evaluation is necessary in newborns and children, regardless of the degree of hearing loss, especially if there is a family history of hearing and vision impairment. For the long-term success of the rehabilitation process of individuals with Usher syndrome, it is recommended to use the necessary rehabilitative methods together with hearing aids and CI before the loss of two basic senses (hearing and vision).

Koleilat, A., et al. (2020). **Current approaches to the management of Usher syndrome for the clinician**. *Perspectives on the ASHA Special Interest Groups 08-01-Aug2020*, 5(4), 907-916. [Request full text](#)

The purpose of this review article is to advance the understanding of the clinical symptoms of an individual with usher syndrome and highlight the latest research in the development of new treatments including gene therapy. The care and management of individuals with Usher syndrome required a team of multidisciplinary health care professionals, including and audiologist, a speech-language pathologist, an ophthalmologist, and a medical geneticist. It highlights the importance of

early identification for better speech and developmental outcomes, specifically in pediatric individuals.

APPENDIX

SEARCH METHODOLOGY

A systematic search was conducted for literature. The results were screened by librarians using [Covidence](#).

SEARCH LIMITS

- English-language
- Published within the last 10 years

DATABASES SEARCHED

- Medline – index of peer reviewed articles across health sciences and medicine.
- Embase – index of biomed and pharmacological peer reviewed journal articles.
- Emcare – index of nursing, allied health, critical-care medicine and more.
- CINAHL – index of nursing publications.
- Scopus
- Grey literature – Google, Google Scholar, Trip database, Biomed Central Proceedings.

ADDITIONAL SEARCHING

- Forwards and backward citation searching was undertaken for following articles.
 - Saihan, Z., et al. (2009). **Update on Usher syndrome**. *Current Opinion in Neurology*, 22(1), 19–27. <https://doi.org/10.1097/wco.0b013e3283218807>
 - Koleilat, A., et al. (2020). **Current approaches to the management of Usher syndrome for the clinician**. *Perspectives on the ASHA Special Interest Groups 08-01-Aug2020*, 5(4), 907-916.

MEDLINE SEARCH STRATEGY

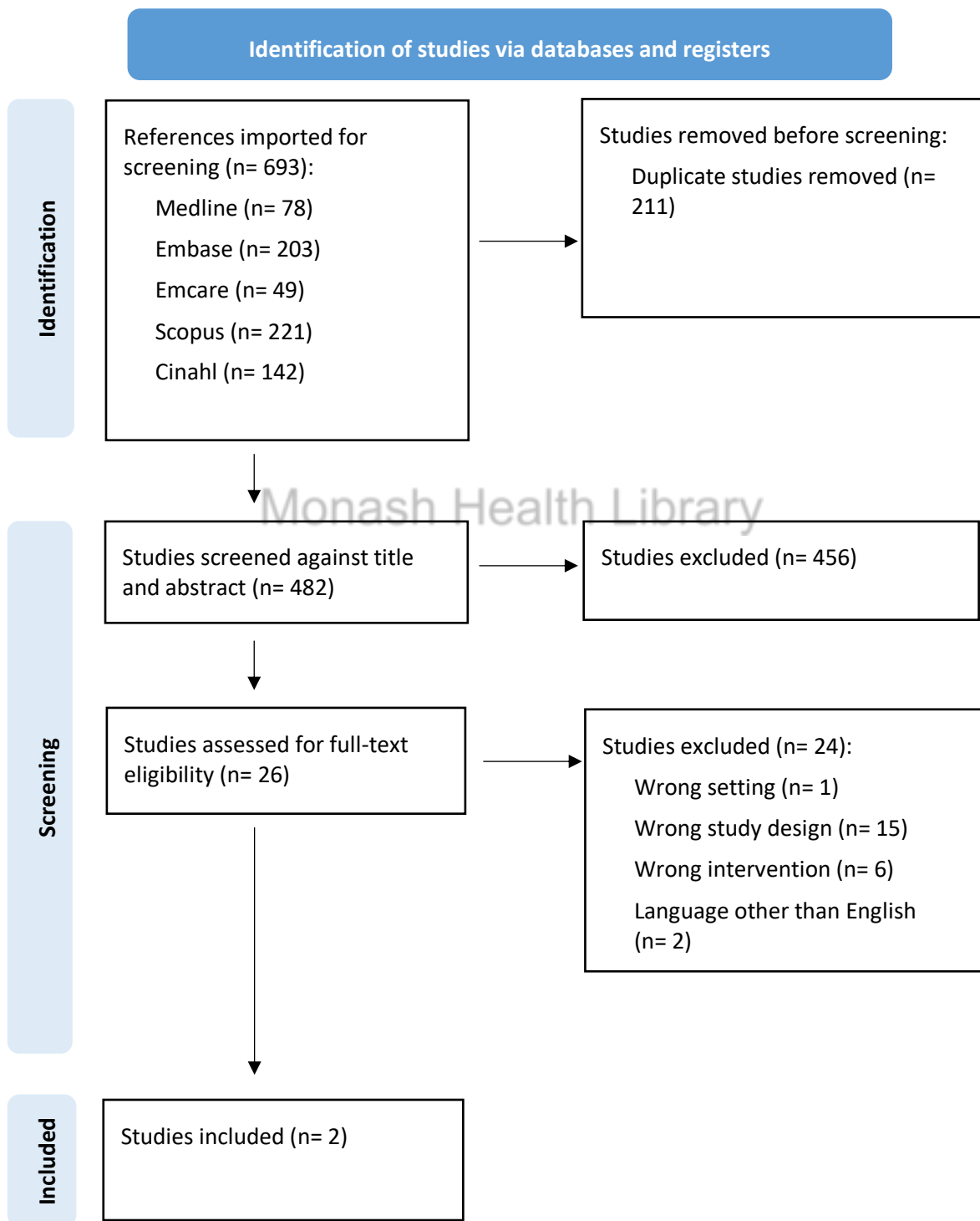
This search strategy was conducted on 18/02/2026 and translated to other databases, as relevant. Searches in each database were conducted on the same day.

Ovid MEDLINE(R) ALL <1946 to February 17, 2026>

- 1 ((audiolog* or audiometr* or auditory or tympanometr* or otologic* or otoscop* or vestibul* or hearing) adj3 (assess* or test* or diagnos* or finding* or surveillance or follow* or follow-up or follow up or program* or screen* or evaluat* or monitor*)).tw,kf. 50962
- 2 Audiologist*.tw,kf. 2408
- 3 Audiology/ or exp Diagnostic Techniques, Otological/ or Audiologists/ 63526
- 4 1 or 2 or 3 96261
- 5 ((usher or hallgren or usher-hallgren or retinitis pigmentosa-dysacusis or dystrophia retinae dysacusis) adj syndrome).tw,kf. 1492
- 6 (usher adj (type 1 or type I or type 2 or type ii or type 3 or type iii)).tw,kf. 68

- 7 (USH1 or USH2 or USH3).tw,kf. 352
- 8 5 or 6 or 7 1514
- 9 4 and 8 182
- 10 limit 9 to (english language and last 10 years) 78

PRISMA CHART



This report contains curated literature results against a unique set of criteria at a particular point in time. Users of this service are responsible for independently appraising the quality, reliability, and applicability of the evidence cited. We strongly recommend consulting the original sources and seeking further expert advice.

Monash Health Library