The Clinical Picture of Large Vestibular Aqueduct Syndrome
Kristen L. Janky, Au.D., Ph.D., M. Geraldine Zuniga, M.D., and C. Matthew Stewart, M.D., Ph.D.

1. LVAS stands for:
   a. Long Vestibular Aqueduct Syndrome
   b. Large Vestibular Aqueduct Syndrome
   c. Large Vestibular Aqueduct Synapsis
   d. Large Vertical Aqueduct Syndrome

2. True or False:
   LVAS is one of the most common congenital inner ear malformations.
   a. True
   b. False

3. The following tests should be done to assist in the diagnosis of LVAS:
   a. CT & MRI
   b. CT & traditional audiometry including bone conduction audiometry
   c. Tympanometry & acoustic reflex testing
   d. All of the above
   e. None of the above
   f. A & C

4. The typical presentation of LVAS is that of down-sloping hearing loss, often accompanied by a conductive component, with progressions in hearing loss occurring either spontaneously or paired with a precipitating event.
   a. True
   b. False

5. LVAS presents a varied clinical picture in terms of:
   a. Hearing loss configuration and type
   b. Onset and severity of hearing loss
   c. Incidence of progression
   d. Presence of vestibular complaints
   e. It’s association with other inner ear anomalies
   f. A, B, & C
   g. All of the above
   h. A, C & D
6. The Vestibular Aqueduct is described by Valvassori and Clemis as:
   a. A J-shaped bony canal coursing from the vestibule to the anterior portion of the petrous pyramids and houses the endolymphatic duct and sac
   b. A J-shaped bony canal coursing from the vestibule to the posterior portion of the petrous pyramids and houses the endolymphatic duct.
   c. A G-shaped bony canal coursing from the vestibule to the posterior portion of the petrous pyramids and houses the endolymphatic duct and sac
   d. A J-shaped bony canal coursing from the vestibule to the posterior portion of the petrous pyramids and houses the endolymphatic duct and sac

7. The VA is generally considered enlarged when the diameter is greater than 0.5 mm
   a. True
   b. False

8. Some individuals have been documented as having LVAS with normal hearing.
   a. True
   b. False

9. Which statement describes the classic depiction of hearing loss with LVAS the best:
   a. Congenital, sloping hearing loss (greater in the high frequencies) with eventual progression.
   b. Congenital hearing loss in the low frequencies with eventual progression.
   c. Congenital, sloping hearing loss (greater in the high frequencies) which is consistent over time

10. True or False:
    LVAS more commonly occurs unilaterally and often presents with a symmetrical hearing loss.
    a. True
    b. False

11. Air bone gaps have been reported in as many as ____% of individuals with LVAS
    a. 88
    b. 80
    c. 8
    d. 50
    e. 55
12. The largest air bone gaps are generally documented in the:
   a. Higher frequencies
   b. Lower frequencies
   c. Mid frequencies

13. In the majority of cases, the conductive hearing loss presents with:
   a. Normal tympanometry
   b. Normal reflexes
   c. Normal umbo velocity
   d. A & B
   e. A & B
   f. A, B, & C

14. a. Because there is such a high incidence of sensori-neural and/or mixed hearing loss in LVAS, bone conduction testing should be routinely assessed, regardless of the severity of air conduction thresholds.
   b. Because there is such a high incidence of conductive and/or mixed hearing loss in LVAS, bone conduction testing should be routinely assessed, regardless of the severity of air conduction thresholds.
   c. Because there is such a high incidence of conductive and/or mixed hearing loss in LVAS, bone conduction testing should be routinely assessed, except for mild hearing losses.
   d. Because there is such a low incidence of conductive and/or mixed hearing loss in LVAS, bone conduction testing does not need to be routinely assessed, regardless of the severity of air conduction thresholds.

15. Although it’s often omitted in audiometric testing, bone conduction testing at ____ Hz may be especially useful in this population.
   a. 250
   b. 500
   c. 1000
   d. 2000
   e. 4000

16. Syndromes with which LVAS can be associated include:
   a. Pendred syndrome
   b. Distal renal tubular acidosis
   c. Branchio-oto-renal syndrome
   d. Waardenburg syndrome
   e. X-linked congenital mixed deafness
   f. CHARGE syndrome
   g. All of the above
   h. A, B & E
17. Cochlear implants have been a beneficial treatment when hearing loss has progressed beyond the degree of amplification.
   a. True
   b. False

18. Despite a variety of treatments and recommendations that have been attempted, none have proven to ultimately halt or permanently improve hearing in individuals with LVAS.
   a. True
   b. False

19. a. LVAS can be associated with nonsyndromic hereditary hearing loss
    b. LVAS can be associated with DFNB4
    c. LVAS can be associated with syndromic SNHL
    e. LVAS can be associated with nonsyndromic hereditary hearing Loss, DFNB4 and syndromic SNHL

20. Reported symptoms of dizziness and/or vertigo are variable. It can range from no symptoms to mild disequilibrium and episodic true vertigo.
   a. True
   b. False