

## **GEORGIA SENSORY ASSISTANCE PROJECT USHER SYNDROME SCREENING**

Usher Syndrome is an inherited disorder characterized by serious hearing impairment that is present at birth, and a progressive loss of vision caused by Retinitis Pigmentosa (RP). Some children will also have balance problems and be late walkers. Approximately 3% of congenitally deaf persons and over 50% of the deaf-blind community have Usher Syndrome. Usher Syndrome can be difficult to diagnose in young children because the effects of the RP do not show up until later. A reliable diagnosis can usually be obtained around the age of twelve. The Georgia Sensory Assistance Project (GSAP) has developed a vision screening protocol for students who are deaf and hard of hearing that includes screening for Usher Syndrome. For more information on screening for Usher Syndrome and training on the protocol, please contact Martha Veto at GSAP – [mveto@gsu.edu](mailto:mveto@gsu.edu), or 404-413-8312.

### **CHARACTERISTICS OF USHER SYNDROME**

#### **What are behavioral symptoms teachers and parents may see that suggest Usher Syndrome?**

- Congenital hearing loss
- Increasing difficulty seeing in the dark or to the side
- Vestibular problems like poor balance
- Difficult seeing when coming in from bright sunlight
- Bright light hurts eyes
- Startles easily because of restricted fields
- Can't find small objects that have dropped
- Stumbles on stairs or curbs
- Considered clumsy, loses balance easily in the dark

#### **Characteristics that suggest a referral for Usher Syndrome Screening:**

- Only the eyes and ears are affected
- Normal intelligence
- Abnormal balance
- Severe hearing loss
- Early vision problems include night blindness, loss of peripheral fields

#### **Why is it important to diagnose Usher Syndrome as soon as possible?**

- The gradual decrease in vision may go unnoticed by an individual who may continue activities (such as driving) that cannot be safely done anymore.
- Parents and teachers can plan for educational and vocational experiences and guidance that take into account the eventual visual difficulties.
- Diagnosis of Usher Syndrome in an older child allows parents to consider genetic counseling.