VATER/VACTERL ASSOCIATION

What is it?

VATER/VACTERL describes a group of non-random, co-occurring congenital malformations that are linked together under an umbrella title. Each defect is caused by an unknown genetic mutation. Since these malformations were occurring together more often than expected, the condition was termed an association. Infants born with the association are usually small and have further trouble gaining weight. It is sometimes associated with hydrocephalus or agenesis of the corpus callosum, but not always. Some malformations may be diagnosed during prenatal ultrasound or at birth. A VATER/VACTERL diagnosis occurs when at least 3 of the below are present:

V- Vertebra

Vertebral defects occur in about 70% of cases and involve issues of the spinal column like malformed vertebra or vertebral dysplasia. Also commonly accompanied by rib anomalies.

A- Anal

Imperforate anus can be caused by a disconnected passageway with the large intestine, narrowing of the anus or no anus. Anal atresia occurs in about 55% of cases when a thin covering blocks the anal opening. Complications are usually more serious in boys.

C- Cardiac

Cardiac anomalies occur in about 75% of patients. The most common defect is ventricular septal defect but can also include arterial septal defect or tetralogy of Fallot. The location and size of the defect predicts the severity of symptoms. Small defects may close on their own or as the child grows. In more serious defects, the heart cannot properly pump blood to the lungs and body leading to congestive heart failure.

TE- Tracheoesophageal

Tracheoesophageal fistula and/or atresia occurs in about 70% of patients. With TE atresia, the passage way between the esophagus and the stomach narrows or ends in a pouch, disabling food from entering the stomach. With a TE fistula, there is an abnormal connection between the tracheae and the stomach which can cause food to be aspirated into the lung.

R- Renal

Renal or kidney anomalies occur in about 50% of cases. Defects include incomplete formation of one kidney and/or urologic complications like obstruction of urine, urine reflex, or inability to empty. Kidney failure may also result.

L: Limb

Limb anomalies include structural issues with the forearm on the radial side. Sometimes, the radial bone does not grow. Other times, the thumb does not develop and/or an extra bone in the thumb develops. Other effects include extra digits, webbed fingers or club foot.

What is the treatment?

Treatment is focused on the specific malformations and the related symptoms. A team of medical and developmental specialists are key to the best possible outcome. Those diagnosed with VACTERL Association will often be followed by numerous specialists which might include cardiologists, urologists, orthopedists, ear nose and throat physicians and clinical geneticists. The pediatrician will often manage the multidisciplinary approach. Surgery is often needed to repair the birth defects.

Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

Supporting students with VACTERL Association in the school setting require educators and parents/guardians to work as a team. A well coordinated plan promotes success for the student in the educational setting. Some accommodations to consider for an 504/IEP could include:

- PT/OT/SLP/Vision/Hearing evaluations
- Plans for mobility accommodations
- Plan for absences and make-up work
- Adapted PE considerations to accommodate motor development
- Consider assistive technology
- Presenting materials in various formats for the student
- Consider extra adult support
- Organizational strategies and aids
- Flash pass to exit room or use restroom
- Offer designated rest area
- Offer preferential seating
- Emotional support
- Behavioral support
- Staff education and training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including all affected systems and symptoms
- Current medication list for school and home
- Communicate with school staff, parents, and provider any changes or concerns about the disease
- Shunt information, if applicable, including emergency protocol for shunt
- Nutrition orders and/or feeding protocol including snacks and oral fluids
- Feeding tube orders including flush and replacement per county policy, if applicable
- Catheterization orders including catheter type and size, times, position
- Colostomy orders including supplies and skin care, if applicable
- Rest breaks
- Baseline cardiac assessment with parameters
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

National Organization for Rare Diseases

https://rarediseases.org/rare-diseases/vacterl-association/

VACTERL Association-Genetic and Rare Diseases Information Center

https://rarediseases.info.nih.gov/diseases/5443/vacterl-association

VACTERL Association-Genetics Home Reference

https://ghr.nlm.nih.gov/condition/vacterl-association