Factsheet: 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 also 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 association 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.
**Suggested school accommodations**

- PT/OT/SLP/Vision/Hearing consultations
- Plans for mobility accommodations
- Adaptive PE considerations to accommodate motor development
- Presenting materials in various formats for the student
- Organizational strategies and aids
- Assistive technology
- 1:1 support as needed
- Flash pass to exit room or use restroom
- Plans for missed schooling, flexible schedule
- Know behavior signs for when break is needed
- Offer designated rest area
- Offer preferential seating
- Emotional support

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<tr>
<th>SHNIC school nurses information:</th>
<th>Specific health issues for individual health care plans</th>
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<tbody>
<tr>
<td><strong>Diagnosis including all affected areas and symptoms</strong></td>
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<td><strong>Current medication list for school and home</strong></td>
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<td><strong>Baseline cardiac assessment with parameters</strong></td>
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<td><strong>Shunt information, if applicable, including emergency protocol for shunt</strong></td>
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<td><strong>Nutrition orders and/or feeding protocol including snacks and fluids</strong></td>
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<td><strong>Feeding tube orders including flush and replacement, if applicable</strong></td>
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<td><strong>Catheterization orders including catheter type and size, times, position</strong></td>
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<td><strong>Colostomy orders including supplies and skin care, if applicable</strong></td>
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<td><strong>Rest breaks</strong></td>
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<td><strong>Aid or supervision required</strong></td>
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