

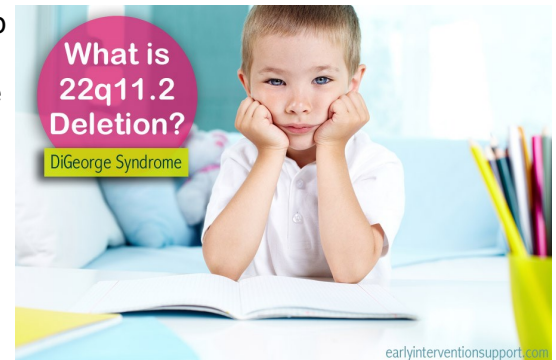
What is it?

DiGeorge syndrome (DGS) is a particular group of clinical features that frequently occur together as a result of a chromosomal 22 defect. The syndrome is normally noticeable right at birth.

Researchers observed a combination of the lack of the thymus gland (immune system) and a lack of parathyroid glands (low calcium in the blood), as well as certain forms of heart disease, developmental delay and behavioral disorders.

Similar disorders including velo-cardiofacial syndrome (VCFS or Shprintzen syndrome) and conotruncal anomaly face (CTAF) syndrome.

The term “22q11.2 deletion syndrome” is commonly used today. DGS results in susceptibility to infection and immune system problems as well as altered facial characteristics, abnormal gland development and even defects in organs like the heart. The severity of the syndrome and the organs affected can range.



What are the symptoms/complications?

Characteristic facial features

- Long face
- Low set, malformed ears
- Underdeveloped chin
- Bulbous nose tip
- Heavy eyelids
- Small mouth, teeth
- Retrognathia (overbite)
- Hypertelorism (abnormal distance between body parts; normally eyes)
- Feeding disorders, dysphagia
- Cleft palate
- Speech issues
- Frequent infections
- Low calcium levels
- Heart defects (pulmonary stenosis, ventricular septal defect, overriding aorta, thick right ventricle)
- Short stature, growth hormone deficiency
- Bone and muscle issues— hypotonia, scoliosis, arthritis
- Renal, pulmonary, gastrointestinal, and ophthalmologic abnormalities
- Higher rates of behavioral, psychiatric, and communication disorders.
 - AD/ADHD
 - Anxiety
 - Autism
 - Affective disorders

Suggested school accommodations

- Consider 504 or IEP
- Strategies for specific learning issues if identified
- Preferential seating due to hearing and attention issues
- Concise directions
- Reduction of stressors
- Check for understanding
- Watch for signs of fatigue
- Breaks
- Various strategies for teaching math
- Use of manipulatives
- Copies of board work
- Visual/Spatial strategies such as more spacing on pages
- Graphic organizers
- Emotional support as many children have difficulty making friends
- Functional Behavioral Plan if behaviors are interfering with learning

SHNIC school nurses information:

Specific health issues for individual health care plans

- Complete diagnosis including baseline assessments for any cardiac, respiratory, kidney, GI issues, feeding issues, immune issues, and psychiatric concerns
- Current medication list; known side effects
- Seizure action plan, if applicable for low calcium levels
- Nutrition orders and feeding protocol if there is a cleft palate and identified feeding disorder
- Note concerns for hearing loss
- Note concerns for behavior and/or supervision
- Allow for rest periods if needed due to cardiac condition and fatigue
- Training for staff regarding Di George syndrome pertinent to student

Resources & Manuals

Mayo Clinic: DiGeorge Syndrome

<http://www.mayoclinic.org/diseases-conditions/digeorge-syndrome/basics/definition/con-20031464>

Merck Manual: DiGeorge Syndrome

<http://www.merckmanuals.com/professional/immunology;-allergic-disorders/immunodeficiency-disorders/digeorge-syndrome>

Children's Hospital of Wisconsin: DiGeorge Syndrome

<http://www.chw.org/medical-care/endocrine/endocrine-conditions/disorders-affecting-calcium-metabolism/digeorge-syndrome/>

Dempster Family Foundation– Pitching in for 22q

<http://dempsterfamilyfoundation.org/living-with-22q/22q-stories>
(School resource and teacher section)