Down syndrome is a chromosome disorder associated with an extra chromosome (Trisomy 21) resulting in intellectual disability and specific physical features. (ICD-9 code 758)

**Prevalence**
- < 1%
- Three types of chromosomal abnormalities can lead to Down syndrome:
  - Nondisjunction – 95% (males 59%, females 41%)
  - Translocation – 4% (females 74%, males 26%)
  - Mosaicism – 1% (may have more subtle features)
- Medical complications seem to be similar in all three groups

**Manifestations**

**Clinical**
- Increased risk of abnormalities in almost every organ system:
  - Intellectual disability and delayed growth
  - Vision and hearing problems
  - Cardiac defects (VSD, ASD, PDA, Tetralogy of Fallot)
  - Characteristic physical features: brachycephalic skull, prominent epicanthic skin folds, small low-set ears, reduced muscle tone, pelvic dysplasia, transverse palmar crease, broad hands and feet, short fingers, and lenticular opacities

**Oral**
- Early onset severe periodontal disease (most significant oral health problem)
- Lower prevalence of dental caries
- Delayed eruption of permanent teeth, malocclusion
- Congenitally missing and malformed teeth are common
- Hypoplasia of mid-facial region
- Hypodontia, microdontia
- Macroglossia, fissured and protruding tongue
- Tongue thrust, bruxism, clenching, mouth breathing

**Other Potential Disorders/Concerns**
- Epilepsy
- Cardiac defects
- Atlantoaxial instability (fragility of cervical vertebrae/spinal cord)
- Compromised immune system
- Sleep apnea
- Increased risk of leukemia
- Hearing loss
- Vision problems
- Hypothyroidism

**Management**

**Medication**
- Medications are prescribed based on symptoms:
**Behavioral**

Many children with Down syndrome can successfully be treated in the dental office.

**Guidance:**
- Plan a pre-appointment (in person/phone) to discuss patient special needs prior to the first visit. Discuss this with the parent or care provider—they know the child best.
- Schedule appointments early in the morning or best time of day for patient.
- Talk with the parent or caregiver to determine the patient's level of intellectual and functional abilities and explain each procedure at a level the patient can understand.
- Use short, clear instructions and speak directly to the patient.
- Minimize distractions, such as sights and sounds, which may make it difficult for the patient to cooperate.
- Start the oral examination slowly, using only fingers at first. If this is successful, begin using dental instruments.
- Use the Tell-Show-Do approach when introducing new instruments or procedures.
- Reward cooperative behavior with positive verbal reinforcement.
- Develop trust and consistency between the dental staff and the patient. Use the same staff, dental operatory, and appointment time each visit if appropriate.

**Dental Treatment and Prevention:**
- Consider patient's cardiac status and need for premedication—medical consult may be indicated.
- It is not uncommon to encounter patients who are tube-fed among the population of Children with Special Healthcare Needs. Patients fed by tube typically have low caries, rapid accumulation of calculus, GERD (Gastro-esophageal Reflux Disease), oral hypersensitivity, and are at high risk for aspiration in the dental chair. No antibiotic premedication is needed for Gastric or Nasogastric tubes. Position the patient in as upright a position as possible and utilize low amounts of water and high volume suction to minimize aspiration.
- Examine patients by the first birthday; monitor tooth eruption patterns and malformations.
- Monitor periodontal disease. Treat as needed and consider specialty referral if indicated.
- Powered toothbrushes may be too stimulating for some children and should be recommended only after determining if the child will tolerate one.
- Consider prescribing Chlorhexidine or other antimicrobial agents for daily use.
- Some patients are good candidates for full orthodontic treatment. Maintain primary teeth as long as possible and consider space maintenance and orthodontic consultation for missing teeth.

**Considerations due to associated medical conditions:**
- Increased gag reflex during oral examination.
- Chronic respiratory infections and open mouth posture → frequent mouth breathing → xerostomia.
- Seizure management during treatment: Remove all dental instruments from the mouth. Clear the area around the dental chair. Stay with the child and turn child to one side. Monitor airway to reduce risk of aspiration. Note time seizure begins: if seizure continues >3 min call EMS – Danger of Status Epilepticus (potentially life threatening).
- Carefully move patients with atlantoaxial instability into the dental chair, giving special attention to the spine and neck. Use pillows to stabilize the patient and increase comfort, as directed by the caregiver.
Look for signs of physical abuse during the examination. Note findings in chart and report any suspected abuse to Child Protective Services, as required by law. Abuse is more common in children with developmental disabilities and often manifests in oral trauma.

Additional information: Special Needs Fact Sheets for Providers and Caregivers

References

Additional Resources
- NIH Institute for Down Syndrome
- Special Care: an Oral Health Professionals Guide to Serving Young Children with Special Health Care Needs
- Bright Futures Oral Health Pocket Guide
- MCH Resource Center
- ASTDD-Special Needs
- NOHIC-NIDCR publications
- Free of charge CDE courses: MCH Oral Health CDE (4 CDE hours); NIDCR CDE (2 CDE hours)