Prevalence
Approximately 1%

Manifestations

Clinical
varies with type of congenital defect (e.g. atrial/ventricular septal defects, pulmonary/aortic stenosis, transposition, heart valve abnormalities)
• Pulmonary congestion/labored breathing
• Heart murmur
• Cardiomegaly
• Congestive heart failure
• Hypoxic spells
• Cyanosis
• Poor physical development
• Clubbing of the terminal phalanges of the fingers

Oral
• Infective endocarditis risk from dental treatment
• Post-operative bleeding risk in children with anti-coagulated status following surgical procedures
• May have oral manifestations caused by co-occurring disorders

Other Potential Disorders/Concerns
• Depression/Anxiety
• Genetic and syndromic conditions (~11%) such as Down, Turner, Marfan and Ehler Danlos syndromes; osteogenesis imperfecta
• Asthma
• Intellectual disabilities
• Esophageal atresia

Management

<table>
<thead>
<tr>
<th>Medication</th>
<th>SIDE EFFECTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diuretics (Furosemide)</td>
<td>Xerostomia. Do not use with aspirin.</td>
</tr>
<tr>
<td>ACE inhibitors (Captopril)</td>
<td>Xerostomia, dysgeusia. Do not use with aspirin.</td>
</tr>
<tr>
<td>Digoxin</td>
<td>Use caution with antibiotics (erythromycin, tetracycline). Consult cardiologist</td>
</tr>
<tr>
<td>Anti-coagulants</td>
<td>Obtain current INR prior to surgical procedures and extractions. Consult with cardiologist to determine if appropriate to pause anti-coagulant therapy or proceed</td>
</tr>
</tbody>
</table>
Behavioral

- Use of nitrous oxide or conscious sedation may be beneficial in reducing anxiety.

Dental Treatment and Prevention

- Obtain thorough medical and dental history at each visit due to risk of infective endocarditis.
- Discuss dental treatment with patient's primary physician or cardiologist. Cardiologist will indicate specific antibiotic prophylaxis needed before dental treatment.
- According to the most recent AHA guidelines, many patients who previously required antibiotic prophylaxis no longer require it. Before discontinuing prophylactic regimen, consult with cardiologist, as some may still recommend prophylaxis.

General Guidelines: Antibiotic Prophylaxis

- Administer a single dose of antibiotic regimen 30-60 minutes before dental procedure.
- Dosage may also be administered up to two hours after procedure if not administered before only in cases when antibiotics are inadvertently not administered.
- Amoxicillin is preferred oral therapy (50 mg/kg). If allergic, consider use of Clindamycin (20 mg/kg), Cephalexin (50 mg/kg), or Azithromycin/Clarithromycin (15 mg/kg).
- Antibiotic prophylaxis recommended for following conditions:
  * High Risk includes prosthetic cardiac valves, previous infective endocarditis, and congenital heart disease (unrepaired cyanotic CHD, including shunts and conduits, completely repaired cardiac defect with prosthetic material or device – for 1st 6 months following surgery, repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or device), cardiac transplant patients who develop valvulopathy)

NOTE: Except for the conditions listed above, antibiotic prophylaxis is no longer recommended for any other form of CHD. If a child has been previously pre-medicated, consult with their physician.

- Antibiotic prophylaxis recommended for following dental procedures:
  * Dental extractions, periodontal procedures, endodontic surgery beyond the apex, dental implant placement and reimplantation of avulsed teeth, initial placement of orthodontic bands, local anesthetic injections, and prophylactic teeth cleaning where bleeding is anticipated.
- Antibiotic prophylaxis is NOT recommended for the following dental procedures:
  * Routine anesthetic injections through non-infected tissue, taking dental radiographs, placement of removable prosthodontic or orthodontic appliances, adjustment of orthodontic appliances, placement of orthodontic brackets, shedding of primary teeth, and bleeding from trauma to the lips or oral mucosa.

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
Children with Congenital Cardiac Disorders continued

References

Additional Resources
- NIH Institute for Congenital Cardiac Disorders
- 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
- Block Oral Disease, MA
- NOHIC-NIDCR publications
- Free of charge CDE courses: MCH Oral Health CDE (4 CDE hours); NIDCR CDE (2 CDE hours)