CHAPTER 11 – CARDIOVASCULAR SYSTEM

First Nations and Inuit Health Branch (FNIHB) Pediatric Clinical Practice Guidelines for Nurses in Primary Care.

The section on Rheumatic Fever (Carditis) has been updated as of December 2017. The remaining content of this chapter was reviewed in September 2011.

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INTRODUCTION

Cardiovascular disease is uncommon in childhood. The major problems seen include congenital heart disease (usually abnormalities of the great vessels, hypoplastic heart, pulmonary or aortic atresia and tetralogy of Fallot), cardiac failure, rheumatic fever carditis and myocarditis. Functional or innocent heart murmurs are common. Congestive heart failure at birth is rare and usually suggests severe valvular deformities.

Symptoms of ventricular septal defect, including heart failure, usually occur at approximately 6 weeks of age. For more information on the history and physical examination of the cardiovascular system in older children and adolescents, see the chapter “Cardiovascular System” in the adult clinical practice guidelines.

ASSESSMENT OF THE CARDIOVASCULAR SYSTEM

Symptoms of cardiovascular disease vary with the age of the child. Ask about:

- Rapid or noisy breathing
- Cough
- Cyanosis
- Sleeping patterns
- Exercise tolerance: indicated in a young child by ability to feed and in an older child by ability to keep up with peers during play

IN INFANTS

CYANOSIS

- An abnormality of oxygen transport related to heart, lungs or blood or inadequate oxygenation of blood due to mixing of venous and arterial blood. Transport problems include impairment of the oxygen-carrying capacity of hemoglobin, as for example, in carbon monoxide poisoning, and hypoxemia secondary to ventilation/perfusion mismatches as for example in pneumonia
- Causes bluish discoloration of mucous membranes, nail beds and skin, is a significant clinical finding and is related to inadequate oxygenation of arterial blood
- May be transient (related to increased oxygen demand by tissues, for example, during feeding in infants or during play in toddlers) or permanent from birth

DIFFICULTY BREATHING

- Tachypnea
- Chest retractions
- Nasal flaring
- Anxious appearance
- Grunting

EXCESSIVE PERSPIRATION

- Infant’s head described as “always wet”
- Infant perspires freely and easily, especially with excretion and feeding

SLOW GROWTH

- Child usually exhibits slow weight gain, relative to height gain; difficulty in feeding may contribute to this problem
- Metabolic demands increased

RESPIRATORY INFECTIONS

- More common with congestive heart failure
- More severe with increased pulmonary flow
IN CHILDREN
– Slow growth
– Respiratory infections
– Chest pain
– Palpitations
– Dizzy spells or blackouts
– Exercise intolerance
– Squatting with cyanotic episodes (“tetralogy spells”)

MEDICAL HISTORY (SPECIFIC TO CARDIOVASCULAR SYSTEM)
– Prematurity (associated with a higher prevalence of congenital cardiac malformation)
– History of illnesses related to heart disease (for example, strep throat)
– “Flu-like” illness
– Joint pains or swelling
– Down syndrome (associated with a higher prevalence of congenital heart disease)

PHYSICAL FINDINGS
An examination of the cardiovascular system involves more than just examining the heart. The examination generally covers two systems: the central cardiovascular system (head, neck and precordium [anterior chest]) and the peripheral vascular system (extremities). Examination of the cardiovascular system must also include a full assessment of the lungs and neuromental status (for signs of confusion, irritability or stupor).

VITAL SIGNS
– Heart rate
– Respiratory rate
– Blood pressure (in both an upper and a lower limb, if possible)
– Temperature (may be elevated with myocarditis or acute rheumatic fever)
– Cardiovascular problems may present as failure to thrive (weight and height below specified percentiles for age) or as a sharp decline in the growth curve across a major percentile line, therefore always document height and weight for all well baby and child examinations

INSPECTION
– Respiratory distress
– Cyanosis: central and peripheral
– Hands and feet: cyanosis, clubbing
– Precordium: visible pulsations
– Edema (hands, feet, sacrum)

PALPATION
– Apical beat is located at fourth intercostal space, lateral to the mid-clavicular line in infants, and at fifth intercostal space, lateral to the mid-clavicular line in older children
– Brief, localized apical tap is normal
– Apical beat may be laterally displaced, which indicates cardiomegaly
– Thrills or heaves may be palpable through chest wall; check supraclavicular area for thrills (in children with a thin chest wall, normal heart movements can be easily palpated and should not be confused with true thrills and heaves)
– Hepatomegaly
– Check for presence, rate, rhythm, amplitude and equivalence of peripheral pulses, especially femoral pulses (which are bounding in patent ductus arteriosus, absent in coarctation of aorta)
– Check for synchrony of radial and femoral pulses
– Capillary refill (normal < 3 seconds)
– Edema: pitting (rated 0 to 4) and level (how far up the feet and legs the edema extends); sacral edema
– Skin: temperature, turgor

AUSCULTATION
– $S_1$ and $S_2$ heart sounds
– Physiologic splitting of $S_2$ heart sound
– Added heart sounds ($S_3$ and $S_4$): determine their location and relation to respiration
– Murmurs: determine location (where murmurs are best heard), radiation, their timing in cardiac cycle, intensity, grade (see Table 1, “Characteristics of Heart Murmurs of Various Grades”) and quality
– Bruits: may occur in carotid arteries, abdominal aorta, renal arteries, iliac arteries, femoral arteries
– Crackles in lungs: may indicate heart failure (in infants and children, this usually occurs as a late sign)
Table 1 – Characteristics of Heart Murmurs of Various Grades

<table>
<thead>
<tr>
<th>Grade</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Very quiet, barely audible</td>
</tr>
<tr>
<td>II</td>
<td>Quiet but audible</td>
</tr>
<tr>
<td>III</td>
<td>Easily heard</td>
</tr>
<tr>
<td>IV</td>
<td>Thrill can be felt, murmur is easily heard</td>
</tr>
<tr>
<td>V</td>
<td>Thrill can be felt and loud murmur can be heard with stethoscope placed lightly on chest</td>
</tr>
<tr>
<td>VI</td>
<td>Thrill can be felt and very loud murmur can be heard with stethoscope held off the chest wall</td>
</tr>
</tbody>
</table>

COMMON PROBLEMS OF THE CARDIOVASCULAR SYSTEM

HEART MURMURS

Most murmurs are innocent flow murmurs, which are present in up to 50% of children; see “Innocent Heart Murmur.”

A heart murmur may signify congenital anatomic, infectious or inflammatory damage to valves and outlets of the four chambers of the heart.

PHYSICAL FINDINGS: AUSCULTATION

Auscultation helps to distinguish significant murmurs from innocent murmurs.

Murmurs must be recognized in relation to other physiologic and pathologic sounds of the cardiac cycle.

- The S₁ sound is caused by the closure of the mitral and tricuspid valves, which usually occurs simultaneously. The S₁ sound is best heard at the cardiac apex.
- The S₂ sound occurs with the closure of the aortic and pulmonary valves. Because the closure of these two valves is somewhat asynchronous, what is known as the S₂ sound actually consists of two sounds. The separation of the two component sounds is often difficult to detect in young children, although it is more pronounced during inspiration. Wide separation of the S₂ sound is often a significant pathologic finding. The S₂ sound is best heard in the second and third left intercostal spaces.
- An S₃ sound may occur after the second heart sound. This may be found in healthy children. It is a sign of heart failure in a symptomatic child. The S₃ sound is best heard when listening at the apex of the heart (in the fourth and fifth intercostal spaces); a left side-lying position may accentuate the sound. Use the bell part of the stethoscope.
- Ejection “clicks” may be present in certain conditions; they are always abnormal.

If a murmur is present, several characteristics should be determined.

Timing within Cardiac Cycle

- Systolic ejection murmurs occur after the first sound. They are caused by turbulence in the blood as it leaves the heart.
- Pansystolic murmurs begin with the first heart sound and end with the second. They most often occur in association with ventricular septal defects.
- Diastolic murmurs begin with the second heart sound. They are always abnormal.

Shape or Contour

- Qualifies the intensity over time: murmurs can be crescendo, decrescendo, or crescendo-decrescendo.
**Location on the Thorax**

There are four general auscultatory areas:

- **Aortic:** left ventricular outflow murmur (usually ejection)
- **Pulmonary:** right ventricular outflow murmur, patent ductus arteriosus
- **Tricuspid:** tricuspid murmurs increase on inspiration; ventricular septal defects are heard best in this area
- **Mitral:** murmur at the cardiac apex

**Radiation**

Radiation of the murmur to the back, sides and neck should be carefully auscultated. Radiation of the murmur may give important diagnostic clues (for example, aortic stenosis radiates to the neck).

**Intensity of Murmur**

- Intensity expressed as a fraction of VI (for example, I/VI, II/VI), where a very loud murmur = V/VI or VI/VI, a loud murmur = III/VI or IV/VI, and a soft murmur = I/VI or II/VI (see Table 1, “Characteristics of Heart Murmurs of Various Grades”)
- Intensity (loudness) does not necessarily correlate with the severity of the condition. Soft murmurs may be dangerous, whereas loud murmurs are not necessarily so. A murmur associated with a thrill has an intensity of at least IV/VI
- Intensity may also increase with increased blood flow, as with exercise

**Pitch**

- Can be low, medium or high and is determined by whether it can be auscultated best with the bell or the diaphragm of a stethoscope

**Quality**

- Blowing
- Harsh
- Musical
- Rumbling
- Clanging

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**INNOCENT HEART MURMUR**

Heart murmur that occurs in the absence of anatomic or physiologic abnormalities of the heart and therefore has no clinical significance. Such murmurs occur in 50–80% of children.

**TYPES OF INNOCENT HEART MURMURS**

Still’s murmur – vibratory, systolic ejection murmur (SEM), lower left sternal border (LLSB) or apex; ages 3 to 6 years.

Venous hum – infraclavicular hum, continuous, heard on right side more than left side; ages 3 to 6 years.

Peripheral pulmonic stenosis – pulmonic area, systolic, low pitched, radiates to axilla and back, seen in neonates; disappears usually by 3 to 6 months of age.

Pulmonary ejection – soft, blowing, upper left sternal border, systolic ejection murmur (SEM).

**PATHOPHYSIOLOGY**

Most innocent heart murmurs are produced by the forward flow of blood, which creates turbulence in the chambers of the heart or the great vessels. These murmurs are often more pronounced in high-output states, such as during a fever. Because the intensity of the murmur parallels the ejection velocity of blood from the ventricles, innocent murmurs usually occur during early to mid-systole, are short in duration, have a crescendo-decrescendo contour (especially an ejection murmur), are less than 3/6 in intensity and are never diastolic.

**CLINICAL FEATURES**

Innocent heart murmurs are asymptomatic and are usually found on routine physical examination.

**DIAGNOSTIC TESTS**

- Electrocardiogram (ECG)
- Echocardiography (only as ordered by a physician)

**MANAGEMENT**

Reassure the parents or caregiver that no immediate treatment is necessary.

**Referral**

Refer the asymptomatic child electively to a physician for assessment when a murmur is found.
EMERGENCY PROBLEMS OF THE CARDIOVASCULAR SYSTEM

CARDIAC FAILURE

The inability of the heart to pump blood commensurate with the body’s needs. The symptoms and signs correlate with the degree of failure.

CAUSES

- Congenital abnormality of cardiac structures
- Inflammatory (for example, rheumatic fever)
- Infectious (for example, viral cardiomyopathy, subacute bacterial endocarditis)
- Severe anemia (that is, hemoglobin < 40 g/L)
- Other high-output states (for example, thyrotoxicosis, arteriovenous malformation)
- Extracardiac disease (for example, chronic pulmonary disease, pulmonary hypertension)

HISTORY

The history varies according to the child’s age.

- Difficulty with feeding
- Shortness of breath
- Excessive sweating
- Poor weight gain
- Anxious appearance

PHYSICAL FINDINGS

- Tachycardia
- Tachypnea
- Blood pressure (assessed in both arms) usually normal but may be reduced (if so, this is cause for concern, as it may indicate cardiogenic shock)
- Temperature: if higher than normal, consider inflammatory or infectious cause
- Irritable
- Anxious
- Fontanel full
- Nostrils flared
- Cyanosis
- Peripheral swelling (in older children)
- Increased jugular venous distention
- Displaced, diffuse apical impulse (cardiomegaly)
- Heave or thrill
- Gallop rhythm (with extra S₃ heart sound)
- Increased murmurs
- Crackles in lung fields
- Hepatomegaly
- Diminished peripheral pulses

DIFFERENTIAL DIAGNOSIS

- Respiratory disease (for example, bronchiolitis or pneumonia)
- Metabolic abnormality (for example, hypoglycemia; poisoning, as with salicylates) also consider hyperglycemia with ketosis, head injuries
- Sepsis including meningitis

COMPLICATIONS

- Decreased cardiac output (shock)
- Death

DIAGNOSTIC TESTS

- Pulse oximetry

MANAGEMENT

Goals of Treatment

- Improve hemodynamic function
- Prevent complications

Appropriate Consultation

Consult with a physician regarding emergency treatment.

Nonpharmacologic Interventions

- Nurse the child in head-elevated position (do not allow neck to become kinked)
- Restrict oral fluids to no more than the quantity required to maintain hydration (see chapter “Fluid Management”)

Adjuvant Therapy

- Start IV therapy with normal saline to keep vein open (unless this would stress the child too much)
- Give oxygen 6–10 L/min or more by non-rebreather mask. Titrate to keep oxygen saturations > 97%
**Pharmacologic Interventions**

Drugs used to treat heart failure in children are to be ordered by a physician.

- Diuretics to decrease volume: furosemide (Lasix), 1 mg/kg IV stat (may be given PO if IV access not available)
- ACE inhibitors may be prescribed by a physician for afterload reduction
- Digoxin may be used in some cases to increase contractility

**Monitoring and Follow-Up**

**Acute Phase**

Monitor ABCs (airway, breathing and circulation), vital signs, pulse oximetry (if available), heart and lung sounds, intake and output until child is transferred to hospital.

**Over the Long Term**

Children with cardiac illness should be monitored regularly within the community to ensure normal growth and development and to watch for complications. Frequency of follow-up depends on the severity of the condition.

**Referral**

Medevac immediately.

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**CYANOSIS IN THE NEWBORN (BIRTH TO 6 WEEKS)**

Bluish discoloration of the skin and mucous membranes secondary to hypoxia.

**CAUSES**

**Congenital Heart Disease**

Cardiac cyanosis is due to left-to-right shunting, so that systemic venous blood bypasses the pulmonary circulation and enters the arterial systemic circulation.

Findings of increased risk for congenital heart disease:

- Genetic syndromes (for example, Down syndrome)
- Certain extracardiac anomalies (for example, omphalocele)
- Maternal diabetes that is poorly controlled in the first trimester
- Exposure to a cardiac teratogen (for example, lithium, isotretinoin [Accutane], alcohol)
- Family history of significant congenital heart disease

**Non-cardiac Causes**

- Pulmonary infection (for example, group B streptococcal infection)
- Intrauterine infection or systemic viral infection (for example, Rubella or Coxsackie B5)
- Aspiration of meconium
- Pulmonary hypoplasia
- Respiratory distress syndrome (for example, in premature infants)
- Hypoventilation (for example, neurologic depression)
- Persistent fetal circulation: seen in post-term infants with perinatal distress or pulmonary disease

**Clinical Features of Infants with Cyanotic Heart Disease**

The clinical features usually present in the first week of life but may present later:

- Difficulty feeding; infant appears to tire easily
- Lethargy
- Cyanosis when feeding or active (for example, while crying)
- Perspiration on face or forehead, especially when feeding or active
- Rapid, noisy breathing

**PHYSICAL FINDINGS**

- Lethargy
- Cyanosis, initially of the oral mucosa; in severe cases, the cyanosis becomes generalized
- Reduced oxygen saturation
- Tachypnea
- Poor perfusion (for example, pallor or gray, ashen appearance; extremities cool; capillary refill diminished; peripheral pulses diminished)
- In coarctation of aorta, pulse quality and blood pressure may differ in different extremities
- Heart sounds may be loud
- Precordium may appear hyperdynamic (heaves or thrills may be present)
- Heart murmur may be present
- Hepatomegaly (if infant is in heart failure)

**DIFFERENTIAL DIAGNOSIS**

- Pulmonary causes as listed above
- Sepsis
COMPLICATIONS
- Cardiac failure (see “Cardiac Failure”)
- Failure to thrive (see “Failure to Thrive” in the pediatric chapter “Hematology, Endocrinology, Metabolism and Immunology”)
- Death

DIAGNOSTIC TESTS
- Pulse oximetry

MANAGEMENT

Appropriate Consultation
Consult a physician immediately and prepare to medevac.

Adjuvant Therapy
- Give oxygen 6–10 L/min (or more, if necessary) by non-rebreather mask. Titrate to keep oxygen saturations > 97%2
- Consider intravenous (IV) therapy with normal saline if infant is feeding poorly or is in significant clinical distress

Nonpharmacologic Interventions
- Nurse in an upright position
- Feed small amounts frequently

Monitoring and Follow-Up
- Monitor level of consciousness, vital signs, heart and lung sounds, perfusion, pulse oximetry
- Hydration status (intake and output) (see “Clinical Features of Dehydration” in the chapter “Fluid Management”)
- Watch for signs of cardiac failure (see “Cardiac Failure”)

Referral
- Medevac as soon as possible
Acute rheumatic fever (ARF) is a diffuse inflammatory disease of the connective tissues involving the heart, joints, skin, central nervous system and subcutaneous tissue. ARF is the immunologic sequela to untreated Group A Streptococcal (GAS) infection. While acute rheumatic fever leaves no lasting damage to the brain, joints or skin, damage to heart valves may be permanent which lead to a chronic heart condition called rheumatic heart disease (RHD). In the absence of secondary antibiotic prophylaxis, recurrence of rheumatic fever is likely and each subsequent episode may cause further cardiac damage to the valves.

**CAUSES**

Untreated GAS pharyngitis infection

**ASSESSMENT**

Medication review: Review current medications, including over-the-counter, complementary and alternative medicines, as well as any chemical or substance intake that may impact management.

Allergy history: Screen for medication, latex, environmental or other allergies and determine approximately when and what type of reaction occurred.

**RISK FACTORS**

- Personal and family history of acute rheumatic fever; most common in those aged 5 to 15 years, but can occur in children as young as 3 years of age

<table>
<thead>
<tr>
<th><strong>OVERVIEW</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Please refer to provincial/territorial guidelines for Acute Rheumatic Fever where available</td>
</tr>
<tr>
<td>Acute rheumatic fever is a serious illness that may require medical evacuation. Consult with physician/nurse practitioner immediately when there is suspicion of acute rheumatic fever.</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>HISTORY OF PRESENT ILLNESS</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Review risk factors and collect history of present illness.</td>
</tr>
</tbody>
</table>

- Preceding GAS pharyngitis
- Joint pain, redness and swelling (migratory arthritis, typically involving the large joints)
- Signs and symptoms of heart failure include:
  - Exertional dyspnea and/or dyspnea at rest
  - Orthopnea
  - Chest pain/pressure and palpitations
  - Cough
  - Fatigue and weakness
  - Nocturia and oliguria
  - Anorexia, weight loss, nausea
- Involuntary, uncoordinated muscular movements (Sydenham’s chorea) (typically presents 2 to 6 months after initial infection)
- Emotional lability
- Non-pruritic, painless rash
- Fever

<table>
<thead>
<tr>
<th><strong>PHYSICAL FINDINGS</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Perform a physical examination using the IPPA approach.</td>
</tr>
</tbody>
</table>

**Major Manifestations**

**Carditis**

- Carditis may be clinical or subclinical.
- New or changing heart murmurs. For more information on heart murmurs, see *Heart Murmurs* in Appendix, Section A of this guide.
- Rubs may be audible with inspiration and expiration if disease is associated with pericarditis.
- Muffled heart sounds (consistent with pericardial effusion).
- Tachycardia at rest; may be out of proportion to fever
– Tachypnea, orthopnea, jugular venous distention, crackles, hepatomegaly, a gallop rhythm, edema and swelling of the peripheral extremities if the client is in heart failure. \(^9\)

– Cardiomegaly, pulmonary congestion and other findings consistent with heart failure may be observed on chest x-ray. \(^{10}\)

**Arthritis**

– Large joints are usually affected, especially the knees and ankles. \(^1\)

– Classified as swelling of the joint in the presence of 2 or more of the following: \(^{11}\)
  - Limitation of movement
  - Hot joint
  - Pain in the joint and/or tenderness

**Sydenham’s Chorea**

– Jerky, uncoordinated movements of the extremities that disappear during sleep. \(^1\)
  - Dysphonia and possible emotional lability \(^6\)

– Female predominance \(^7\)

– Can be a standalone criterion for the diagnosis of acute rheumatic fever without additional manifestations \(^1\)

**Subcutaneous Nodules**

– Usually located over a bony prominence or near tendons \(^{11}\)

– 0.5 to 2 cm in diameter, round, firm, occasionally painful protuberances found on extensor surfaces at specific joints including the knees, elbows, and wrists; also seen in the occiput and along the spinous processes of the thoracic and lumbar vertebrae \(^7\)

**Erythema Marginatum**

– Rare and difficult to detect (especially on dark-skinned people) \(^1\)

– An evanescent, pink rash with a pale center and rounded or serpiginous margins \(^7\)

– The rash is usually present on the trunk and proximal extremities; it is almost never on the face \(^7\)

– Blanches with pressure \(^7\)

– Not affected by anti-inflammatory medication \(^1\)

– Rarely seen as the sole major criterion for acute rheumatic fever and should be accompanied by additional major criteria in order to make the diagnosis \(^1\)

**Minor Manifestations**

– Fever \(^7\)

– Arthralgia \(^7\)

– Raised C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) \(^7\)

– Prolonged PR interval on electrocardiogram (ECG) \(^7\)

**DIFFERENTIAL DIAGNOSIS**

Consult physician/nurse practitioner when practice is outside legislated scope and without authorized delegation.

**TABLE 2**

Differential Diagnoses of Arthritis, Carditis and Chorea

<table>
<thead>
<tr>
<th>ARTHRITIS</th>
<th>CARDITIS</th>
<th>CHOREA</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Septic arthritis</td>
<td>- Physiological mitral regurgitation</td>
<td>- Drug intoxication</td>
</tr>
<tr>
<td>- Connective tissue and other autoimmune diseases such as juvenile idiopathic arthritis</td>
<td>- Mitral valve prolapse</td>
<td>- Tic disorder</td>
</tr>
<tr>
<td>- Lyme disease</td>
<td>- Congenital valve disease</td>
<td>- Intracranial tumor</td>
</tr>
<tr>
<td>- Infective endocarditis</td>
<td>- Infective endocarditis</td>
<td>- Lyme disease</td>
</tr>
<tr>
<td></td>
<td>- Myocarditis, viral or idiopathic</td>
<td>- Autoimmune: Systemic lupus erythematosus, systemic vasculitis</td>
</tr>
<tr>
<td></td>
<td>- Kawasaki disease</td>
<td></td>
</tr>
</tbody>
</table>

The diagnosis is based on a collection of signs known as Jones’ criteria; see Table 3, *Revised Jones’ Criteria.*
### COMPLICATIONS

- Rheumatic heart disease due to permanent damage to the heart valves, which leads to valvular disease, cardiac myopathy and sequelae such as:
  - Heart failure
  - Atrial fibrillation
  - Systemic embolism
  - Stroke
  - Endocarditis
  - Need for cardiac surgery
- Recurrent attack, which worsens the damage to the heart
- Death

### TABLE 3
Revised Jones’ Criteria *(7)*

<table>
<thead>
<tr>
<th>MAJOR CRITERIA</th>
<th>MINOR CRITERIA</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Low-risk populations</strong></td>
<td><strong>High-risk populations</strong></td>
</tr>
<tr>
<td>- Carditis (clinical and/or subclinical)</td>
<td>- Carditis (clinical and/or subclinical)</td>
</tr>
<tr>
<td>- Polyarthritis</td>
<td>- Arthritis (monoarthritis or polyarthritis, polyarthralgia)</td>
</tr>
<tr>
<td>- Chorea</td>
<td>- Chorea</td>
</tr>
<tr>
<td>- Erythema marginatum</td>
<td>- Erythema marginatum</td>
</tr>
<tr>
<td>- Subcutaneous nodules</td>
<td>- Subcutaneous nodules</td>
</tr>
</tbody>
</table>

*For all client populations with evidence of preceding group A *Streptococcal* pharyngitis infection:
  - Diagnosis of initial acute rheumatic fever is:
    - 2 major manifestations or 1 major plus 2 minor manifestations
  - Diagnosis of recurrent acute rheumatic fever is:
    - 2 major or 1 major and 2 minor or 3 minor manifestations

**Low-risk populations are those with acute rheumatic fever ≤ 2 per 100,000 school-aged children or all-age rheumatic heart disease prevalence of ≤ 1 per 1,000 population per year.

### DIAGNOSTIC TESTS

Consult physician/nurse practitioner when practice is outside legislated scope and without authorized delegation.

Diagnostic test selection is based on client history, risk factors, physical examination findings and test availability. Testing should be carried out as per provincial/territorial policies and procedures. The tests below are for consideration. Currently, there is no single laboratory test to confirm the diagnosis of ARF.* ARF remains a clinical diagnosis and relies on health professionals being aware of its diagnostic features.*
Laboratory
- Rapid Antigen Detection Test (RADT) (if available)
- Throat culture and sensitivity (C+S) for group A Streptococcus\(^{(10)}\)
- WBC, ESR\(^{(13)}\)
- Blood culture and sensitivity (C+S) if febrile\(^{(13)}\)

Other Diagnostic Tests
- Chest x-ray\(^{(13)}\)
- ECG to assess for prolonged PR interval\(^{(1)}\)

MANAGEMENT

Consult physician/nurse practitioner when practice is outside legislated scope and without authorized delegation.

Note: The diagnosis and treatment of rheumatic fever requires medical evacuation. Emergency treatment of heart failure may be necessary; see FNIHB Pediatric and Adolescent Care Clinical Practice Guidelines – Chapter 11 – Cardiovascular System – Cardiac Failure.

GOALS OF TREATMENT
- Provide symptomatic relief
- Prevent complications

NON-PHARMACOLOGIC INTERVENTIONS

Interventions
- Bed rest while awaiting medical evacuation

PHARMACOLOGICAL INTERVENTIONS

In addition to consulting a physician/nurse practitioner, review the drug monograph, the FNIHB Nursing Station Formulary or provincial/territorial formulary before initiating treatment.

Note: With the exception of acetaminophen or medications to treat heart failure (if required), medications should not be started until the diagnosis has been clearly established. After consultation with receiving facility, antibiotics may be started after throat swab(s) have been collected.\(^{(1)}\)

Antipyretic/Analgesic

Acetaminophen\(^{(4)}\)
- Acetaminophen 10 to 15 mg/kg/dose PO q4-6h PRN
- [caution-hepatic, INR, renal]
- Maximum from all sources: acetaminophen 75 mg/kg/day or 4,000 mg/day, whichever is less

Antibiotic Therapy
- Antibiotics should be initiated to eradicate residual GAS infection in all cases while the diagnosis of ARF is being established.\(^{(1)}\)
  - A full course of antibiotics should be given.\(^{(1)}\)
  - Oral therapy such as penicillin, amoxicillin, cephalaxin or clindamycin may be considered.

For specific dosing recommendations, see FNIHB Pediatric and Adolescent Care Clinical Practice Guidelines – Chapter 9 – Ears, Nose, Throat and Mouth – Bacterial Pharyngotonsillitis – Antibiotic Therapy.

MONITORING AND FOLLOW-UP

Consult physician/nurse practitioner when practice is outside legislated scope and without authorized delegation.

MONITORING
- Monitor vital signs as indicated by client’s condition, including oxygen saturation.
- Monitor intake and output.
- Monitor for signs of cardiac failure. If client is in cardiac failure, see FNIHB Pediatric and Adolescent Care Clinical Practice Guidelines – Chapter 11 – Cardiovascular System – Cardiac Failure.
  - If administering an agent with risk of anaphylaxis, monitor the client closely for 30 minutes.

FOLLOW-UP

Post-acute Phase
- All clients should receive regular follow-up. The frequency and duration of review is dependent on individual clinical needs and should become more frequent in the event of symptom onset, symptomatic deterioration or a change in clinical findings.\(^{(1)}\)
– Discuss the importance of ongoing secondary prophylaxis during every health professional interaction with the client.\(^{(1)}\)
– Because of the risk of recurrence, continual antibiotic prophylaxis with benzathine penicillin G must be maintained.
– The risk of recurrence is greatest in the 5 years after the initial episode.
– Prophylaxis is initiated immediately after completion of a full therapeutic course of antibiotics, as initiated by a physician/nurse practitioner.
– The physician/nurse practitioner should also determine any discontinuation of prophylaxis.
– The duration of antibiotic prophylaxis can range from 5 years to 30 years, as it depends on a number of factors, including age, clinical pattern, environment and time elapsed since the last episode of ARF.\(^{(1)}\)
– Clients may need to be on other cardiac and anticoagulation therapies, depending on the severity of the heart damage due to ARF.

**Referral**
– Arrange for medical evacuation.

**APPENDIX**

**SECTION A: SUPPLEMENTAL CLINICAL MANAGEMENT INFORMATION**

**Heart Murmurs**
– Those most commonly heard during acute rheumatic fever are:\(^{(8)}\)
  • Apical pansystolic murmur is a high-pitched, blowing-quality murmur of mitral regurgitation that radiates to the left axilla. The murmur is unaffected by respiration or position.
  • Apical diastolic murmur (known as Carey-Coombs murmur) is heard with active carditis and accompanies severe mitral insufficiency.
  • Basal diastolic murmur is an early diastolic murmur of aortic regurgitation and is a high-pitched, blowing, decrescendo heard best along the right upper and mid-left sternal border after deep expiration while the client is leaning forward.

**Approaches to Disease Prevention**
Effective control programs for acute rheumatic fever integrate features of primordial, primary and secondary prevention.

**Primordial Prevention**
– Primordial prevention refers to broad social, economic and environmental initiatives undertaken to prevent or limit the impact of GAS infection in a population.

**Primary Prevention**
– Primary prevention refers to medical intervention using antibiotics to reduce GAS transmission, acquisition, colonization and carriage, or treating GAS infection effectively to prevent the development of acute rheumatic fever.\(^{(13)}\)

**Secondary Prevention**
– Secondary prevention refers to medical intervention using long-term prophylactic antibiotics to reduce repeated acquisition of GAS that might induce recurrent episodes of acute rheumatic fever in order to prevent the development of rheumatic heart disease, or for those clients who have established rheumatic heart disease in order to prevent disease progression.

**BIBLIOGRAPHY**
The following references and other sources have informed the updating of this Clinical Practice Guideline.

**REFERENCES**


OTHER SOURCES

Health Canada. First Nations and Inuit Health Branch (FNIHB) Nursing Station Formulary and Drug Classification System. 2016 April.


Canadian Pharmacists Association. Compendium of Therapeutic Choices: Canada’s Trusted Reference for Primary Care Therapeutics (CTC 7). Ottawa: Canadian Pharmacists Association; c2014.


**VIRAL MYOCARDITIS**

Myocarditis is an inflammatory disorder of the myocardium with necrosis of the myocytes and associated inflammatory infiltrate.

**PATHOPHYSIOLOGY**

Myocarditis generally results in a decrease in myocardial function, with concomitant enlargement of the heart and an increase in the end-diastolic volume caused by increased preload. Progressive increase in left ventricular end-diastolic volume increases left atrial, pulmonary venous and arterial pressures, resulting in increasing hydrostatic forces. These increased forces lead to both pulmonary edema and congestive heart failure.

**CAUSES**

It is usually caused by a viral infection. Parvovirus B19 and human herpesvirus-6 are the most frequent pathogens in patients with acute myocarditis. Infecting organisms may include the following:

- Parvovirus B19
- Herpesvirus
- Coxsackievirus types A and B (especially type B)
- Adenovirus (most commonly types 2 and 5)
- Cytomegalovirus
- Echovirus
- Epstein-Barr virus
- Hepatitis C virus
- Human immunodeficiency virus
- Influenza and parainfluenza
- Measles
- Mumps, associated with endocardial fibroelastosis (EFE)
- Poliomyelitis virus
- Rubella
- Varicella

**Risk Factors**

Younger patients, especially newborns and infants and immunocompromised individuals may have increased susceptibility to myocarditis.

**HISTORY**

Clinical presentation varies widely. In mild forms, few or no symptoms are noted. In severe cases, patients may present with acute cardiac decompensation and progress to death.
In newborns and infants, symptoms may sometimes appear suddenly and may include:

- Irritability
- Failure to thrive
- Feeding difficulties
- Fever and other symptoms of infection
- Lethargy
- Low urine output (a sign of decreasing kidney function)
- Pale hands and feet (a sign of poor circulation)
- Rapid breathing
- Rapid heart rate

Symptoms in children over age 2 may also include:

- Belly area pain and nausea
- Cough
- Fatigue
- Swelling (edema) in the legs, feet and face
- Recent, nonspecific, flu-like illness
- Older children present with similar symptoms as above and may experience lack of energy and general malaise
- Chest pain: Although rare in young children, this may be the initial presentation for older children and adolescents and should be considered a serious symptom accordingly

**PHYSICAL FINDINGS**

**Neonates/Infants**

- Hypothermia or hyperthermia
- Tachypnea
- Tachycardia
- Cyanosis
- Cool extremities
- Decreased capillary refill
- Pale or mottled skin may be present
- Wheezing, and diaphoresis with feeding
- Irritability
- Somnolence
- Hypotonia
- Seizures
- Oliguria
- End-organ damage (for example, renal failure) may develop because of direct viral infestation or because of low cardiac output

**Older Children**

- Low grade fever
- Tachycardia, weak pulse
- Jugular venous distention and edema of the lower extremities may be present
- Heart sounds may be muffled, especially in the presence of pericarditis
- An S3 may be present
- Heart murmur caused by atrioventricular valve regurgitation may be heard
- Crackles may be heard in older children
- Hepatomegaly may be present in younger children
- Cool extremities
- Decreased capillary refill
- Pale or mottled skin may be present

**Adolescents**

Presentation in adolescents is similar to that of children between 6 and 12 years old. However, the following symptoms may be more prominent:

- Decreased exercise tolerance
- Lack of energy, malaise
- Chest pain
- Low-grade fever
- Arrhythmia
- Cough
- Low cardiac output

**DIFFERENTIAL DIAGNOSIS**

- Myocarditis, nonviral
- Pericarditis, viral
- Aortic stenosis, valvular
- Enteroviral infections
- Cardiomyopathy, dilated
- Glycogen-storage disease type I or type II
- Coarctation of the aorta
- Coronary artery anomalies

**DIAGNOSTIC TESTS**

Chest x-ray may show cardiomegaly and cardiac failure.

Electrocardiography (ECG)
In some patients with mild cardiac involvement, ECG changes may be the only abnormal findings suggestive of myocarditis.

- Low-voltage QRS (< 5 mm throughout the limb leads) is the classic pattern
- Pseudoinfarction patterns with pathologic Q waves and poor progression of R waves in the precordial leads may also be present
- T-wave flattening or inversion is a common finding associated with small or absent Q waves in V5 and V6
- Left ventricular hypertrophy with strain may be present
- Other nonspecific findings include a prolonged PR segment and prolonged QT interval
- Sinus tachycardia is the most common finding
- Premature ventricular contractions and atrial tachycardia have been reported
- Junctional tachycardia is common and may worsen congestive heart failure
- Occasional second-degree and third-degree atrioventricular block may be present
- Ventricular tachycardia is commonly associated and may be the initial presentation

**COMPLICATIONS**

- Arrhythmia
- Cardiac failure (see “Cardiac Failure”)
- Thromboembolism
- Decrease in ventricular function
- Dilated cardiomyopathy

**MANAGEMENT**

**Goals of Treatment**

- Stabilize cardiovascular function
- Prevent complications

**Adjuvant Therapy**

- Give supplemental oxygen as necessary via non-rebreather mask. Titrate to keep oxygen saturations > 97%
- Start an intravenous line with normal saline. Run at a rate sufficient to maintain hydration depending on oral intake of child. Do not overhydrate. Keep line open until consultation with an emergency physician. Always weigh infant before starting any intravenous fluids as a measure of hydration

**Nonpharmacologic Interventions**

- Bed rest is necessary during the acute phase of the illness
- Nurse in an upright position

**Pharmacologic Interventions**

Consult a physician for medication orders. Medications may include the following, when indicated: see “Cardiac Failure”.

- Diuretics to decrease volume: furosemide (Lasix), 1 mg/kg IV stat (may be given PO if IV access not available)
- ACE inhibitors may be prescribed by a physician for afterload reduction
- Digoxin may be used in some cases to increase contractility
- Antiarrhythmics
- Anticoagulants

**Monitoring and Follow-Up**

**Acute Phase**

Monitor ABCs (airway, breathing and circulation), vital signs, pulse oximetry, heart and lung sounds, neuromental status, intake and output and medication response and adverse effects closely until child is transferred to hospital.

**Over the Long Term**

Children with cardiac illness should be monitored regularly within the community to ensure normal growth and development and to watch for complications. Frequency of follow-up depends on the severity of the condition.

**Referral**

Medevac to a facility with intensive and cardiology care.
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BOOKS AND MONOGRAPHS


JOURNAL ARTICLES


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END NOTES


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