

HTN in pediatric

- Most childhood and adolescent hypertension in the US is due to primary hypertension.
- Diagnosis and treatment of hypertension in children is important for reducing the risk of:
 - Hypertension and cardiovascular disease in adulthood
 - End organ damage (e.g., left ventricular hypertrophy, CKD)

A. Definitions

1. Normal blood pressures during childhood depend on the child's age, sex, height, and weight. Standards are based on upper arm blood pressures (blood pressures in the legs are generally higher than in the arms).
2. Normal systolic and diastolic blood pressures are defined as blood pressure less than the 90th percentile.
3. Prehypertension is between the 90th and 95th percentiles for age.

In children, hypertension (HTN) is defined as blood pressure (BP) greater than 95th percentile for age, gender, and height measured on at least three different occasions. The context of BP measurement (e.g., cuff size, pain, anxiety) is important. Staging of HTN is listed in Table 166.1. Hypertensive emergency is defined as severe BP elevation associated with target organ damage (encephalopathy, heart failure).

TABLE 166.1 Classification of Blood Pressure	
BLOOD PRESSURE CATEGORY	BLOOD PRESSURE PERCENTILE (%)
Normal	<90th
Prehypertension	90th-95th*
Stage 1 hypertension	95th to (99th + 5 mm Hg)
Stage 2 hypertension	>99th + 5 mm Hg

*If 90th % is greater than 120/80, use 120/80 as the lower limit.

B. Classification. Hypertension is divided into categories based on severity and etiology.

1. Hypertension is defined as the average of three separate blood pressures that are greater than the 95th percentile for age.
2. Stage 1 hypertension is defined as systolic or diastolic blood pressures between the 95th and 99th percentiles + 5 mm Hg.
3. Stage 2 hypertension is defined as systolic or diastolic blood pressures > 99th percentile + 5 mm Hg.
4. Malignant hypertension is hypertension associated with evidence of end organ damage, such as retinal hemorrhages, papilledema, seizures, and coronary artery disease (in adults).
5. Essential hypertension is defined as hypertension without a clear etiology.
6. Secondary hypertension is hypertension that has a recognizable cause (e.g., renal parenchymal disease, coarctation of the aorta). Most hypertension in childhood is secondary hypertension.

Classification of hypertension in children [4][49]

	Age < 13 years [4]	Age ≥ 13 years [4]
Normal blood pressure	• BP < 90th percentile for age, height, and sex	• SBP < 120 mm Hg • AND DBP < 80 mm Hg
Elevated blood pressure	• BP 90–95th percentile • OR BP 120/80 mm Hg to < 95th percentile	• SBP 120–129 mm Hg • AND DBP < 80 mm Hg
Stage 1 HTN	• BP ≥ 95th percentile to < 95th percentile + 12 mm Hg • OR SBP 130–139 mm Hg • OR DBP 80–89 mm Hg	• SBP 130–139 mm Hg • OR DBP 80–89 mm Hg
Stage 2 HTN	• BP ≥ 95th percentile + 12–29 mm Hg • OR SBP ≥ 140 mm Hg • OR DBP ≥ 90 mm Hg	• SBP ≥ 140 mm Hg • OR DBP ≥ 90 mm Hg
Acute severe hypertension	• BP ≥ 95th percentile + 30 mm Hg	• SBP ≥ 180 mm Hg • OR DBP ≥ 120 mm Hg

C. Measurement

1. Blood pressure cuff size

- a. **It is critical to use the proper size cuff.** A cuff that is **too small** will give **falsely elevated blood pressures**, and a cuff that is **too large** will give **falsely depressed blood pressures**.
- b. The cuff bladder should measure two-thirds of the length of the arm from the shoulder to the elbow.

2. Position

- a. Blood pressure in infants should be measured in the supine position.
- b. Blood pressure in children and adolescents should be measured in the seated position with the fully exposed right arm resting on a supportive surface at heart level.

Risk factors for hypertension in children [4]

- Risk factors for hypertension in children are similar to adults (see “Etiology of hypertension” and “Secondary hypertension”).
- **Young children with the following conditions are at risk of early onset (< 3 years old) hypertension:**
 - History of prematurity, very low birth weight, or neonatal complications requiring NICU admission
 - Congenital heart disease including coarctation of the aorta (even if corrected)
 - Known (or family history of) renal disease or congenital anomalies of the kidney and urinary tract
 - Recurrent UTIs
 - Malignancy
 - History of transplant (solid organ or bone marrow)
 - Certain systemic illnesses associated with elevated BP (neurofibromatosis type 1, sickle cell disease, tuberous sclerosis)
 - Elevated ICP
 - Use of medications known to elevate BP
- **Children ≥ 3 years of age are additionally at elevated risk if they have:**
 - Obesity
 - Diabetes
 - Sleep-disordered breathing [4]

D. Etiology (Table 11-1). The causes of hypertension vary with the child's age.

1. **In neonates and young infants**, the most common causes include renal artery embolus after umbilical artery catheter placement, coarctation of the aorta, congenital renal disease, and renal artery stenosis.
2. **In children 1–10 years of age**, the most common causes include renal diseases and coarctation of the aorta.
3. **In adolescents**, the most common causes include renal diseases and essential hypertension.

Pediatric HTN has many causes (Table 166.2) that are either **primary** (essential) or **secondary**. **Essential HTN** is the most common cause of HTN in younger children and adolescents. **Obese children are more likely to develop essential HTN.** Secondary HTN should be suspected with younger age and more severely elevated BP. **Renal disease is the most common cause of secondary HTN in children.**

Essential hypertension*	No identifiable cause, but heredity, salt sensitivity, obesity, and stress all may play a role
Renal diseases	- Glomerulonephritis - Reflux nephropathy - Renal dysplasia - Polycystic kidney disease - Renal trauma - Obstructive uropathy - Hemolytic uremic syndrome
Renal vascular lesions	- Renal artery stenosis or embolus - Renal vein thrombosis - Vasculitis - Neurofibromatosis
Cardiac disease	Coarctation of the aorta
Endocrine disorders	- Neuroblastoma - Pheochromocytoma - Congenital adrenal hyperplasia - Hyperthyroidism - Hyperparathyroidism - Hyperaldosteronism - Cushing syndrome
Central nervous system disorders	- Increased intracranial pressure (e.g., hemorrhage, tumor) - Encephalitis - Familial dysautonomia
Drug-related causes	- Corticosteroids - Illicit drugs (e.g., amphetamines, cocaine, PCP) - Anabolic steroids - Cold remedies - Oral contraceptives
Genetic causes	- Liddle syndrome - Gordon syndrome - Apparent mineralocorticoid excess syndrome - Glucocorticoid remediable - Aldosteronism
Miscellaneous causes	- Wilms tumor - Blood pressure cuff too small - Anxiety - Pain - Fractures and orthopedic traction - Hypercalcemia

*Essential hypertension is rare in young children. All children with hypertension should have a careful evaluation to determine the cause of the hypertension.

PCP = phencyclidine.

E. Clinical features. Clinical presentations of hypertension also vary with the child's age. **Some children, like some adults, are asymptomatic.**

1. **Infants** may present with nonspecific signs and symptoms including irritability, vomiting, failure to thrive, seizures, or even CHF if the hypertension is severe.
2. **Children** with malignant hypertension, especially of acute onset, may develop headaches, seizures, and stroke.
3. **Children** with chronic hypertension may have growth retardation and poor school performance.

Most children with HTN have no symptoms. Signs and symptoms associated with severe HTN include encephalopathy (headache, vomiting, seizures), heart failure, stroke, and retinopathy (blurred vision, flame hemorrhage, and cotton wool spots on retinal exam). Neonatal history (low birth weight or use of umbilical artery catheter); family history of HTN, stroke, or heart disease; and dietary history (excessive salt or caffeine, drugs) are important. Additional findings that may suggest specific causes include abdominal bruit (renal artery stenosis); diminished leg pressure and weak femoral pulse (coarctation of aorta); café-au-lait spots (neurofibromatosis-associated renal artery stenosis); flank mass (hydronephrosis, Wilms tumor); tachycardia with flushing and diaphoresis (pheochromocytoma); and truncal obesity, acne, striae, and buffalo hump (Cushing syndrome). Signs of underlying kidney disease may be present.

DIAGNOSTIC STUDIES

In addition to history and physical examination, evaluation of children with confirmed HTN includes:

1. Baseline etiological assessment (urinalysis, electrolytes, blood urea nitrogen, creatinine, and renal ultrasound)
2. Focused studies based on clinical suspicion (e.g., plasma metanephrines, thyroid studies, vascular imaging)
3. Assessment for target organ damage (echocardiogram for left ventricular hypertrophy)
4. Assessment of other cardiovascular risk factors (lipids, fasting glucose, uric acid)

F. Evaluation. Because most children with hypertension have secondary hypertension, the cause of the hypertension is usually identified through a careful history (including family history and birth history), physical examination, and diagnostic testing. Testing is guided by the most likely causes of hypertension on the basis of the child's age and clinical presentation.

1. Physical examination

- a. All children should undergo an assessment of growth and careful, accurate measurements of four-limb blood pressures to evaluate for coarctation of the aorta (see Chapter 8, section III.F). In coarctation, hypertension is usually noted in the right arm with lower blood pressures in the legs.
- b. In children, a careful fundoscopic examination may show retinal hemorrhages, papilledema, or in long-standing hypertension, arteriovenous nicking.
- c. Other important physical findings include signs of CHF, café-au-lait spots as seen in neurofibromatosis, abdominal masses, abdominal bruits, and ambiguous genitalia.

2. Laboratory evaluation and imaging

- a. Initial evaluation should include a complete blood count (CBC), electrolyte panel, blood urea nitrogen (BUN) and creatinine, urinalysis, plasma renin and aldosterone levels, thyroid panel, chest radiograph, and renal ultrasound. A urine microalbumin/creatinine ratio is useful in detecting early glomerular injury from the hypertension as a sign of end organ damage. (Note that the microalbumin/creatinine ratio is a very sensitive test for detecting proteinuria and is used to detect early glomerular proteinuria in hypertension. The protein/creatinine ratio becomes positive when a patient has more overt proteinuria and is therefore an indicator for more severe damage.)
- b. If the initial evaluation is suggestive, further studies may include plasma catecholamines, measurement of plasma and urinary steroids, and echocardiography. Computer tomography (CT) angiography, especially in the setting of discrepant kidney sizes, can be performed to rule out renal artery stenosis.
- c. Ambulatory blood pressure monitoring is very useful to rule out anxiety ("white coat hypertension") before embarking on more expensive or invasive diagnostic testing.

G. Management. The treatment of hypertension should be aimed at curative therapies, whenever possible. In chronic hypertension, the ultimate goal is to maintain the child's blood pressure below the 90th percentile for age.

1. If a specific, treatable cause of hypertension is identified, directed management, such as surgical correction of a coarctation of the aorta, treatment of hyperthyroidism, or removal of a catecholamine-secreting tumor, is performed as a cure.
2. If the child appears to have essential hypertension, the initial approach is conservative through implementation of a diet with no added salt, and if appropriate, weight loss. This approach requires frequent monitoring and encouragement.
3. If the child has underlying renal disease or more significant hypertension (e.g., stage 2), or if conservative hypertension treatment has failed, antihypertensive medications are used.
4. Hypertensive emergencies (e.g., seizures, severe headache, stroke, fundoscopic changes, CHF) require prompt therapy with IV antihypertensives.

TREATMENT

Therapeutic lifestyle changes (diet, exercise) should be initiated for asymptomatic stage 1 HTN without target organ damage or systemic disease. Medication should be started for stage 2 or symptomatic HTN and stage 1 HTN that fails to respond to lifestyle changes over 6-12 months. Calcium channel blockers or angiotensin-converting enzyme inhibitors are the most frequently chosen first-line options in children. Angiotensin receptor blockers, β -blockers, or diuretics are also effective first-line antihypertensive agents. More than one agent may be needed. Hypertensive emergency requires prompt hospitalization and may require parenteral antihypertensive treatment with nicardipine, labetalol, esmolol, or sodium nitroprusside.

PROGNOSIS

Prognosis depends on underlying etiology and BP control. Essential HTN is usually not associated with morbidity at presentation. Left untreated, however, even asymptomatic stage 1 HTN may increase risk for cardiovascular, central nervous system, and renal morbidity in adults.

COARCTATION OF THE AORTA

ETIOLOGY AND EPIDEMIOLOGY

- Coarctation of the aorta occurs in 10% of all congenital heart defects.
- It is almost always **juxtaductal** in position.
- During development of the aortic arch, the area near the insertion of the ductus arteriosus fails to develop correctly, resulting in a narrowing of the aortic lumen.

CLINICAL MANIFESTATIONS

- Timing of presentation depends primarily on the severity of obstruction and associated cardiac defects.
- Infants presenting with coarctation of the aorta frequently have **hypoplastic aortic arches**, **abnormal aortic valves**, and **VSDs**.
- They may be **dependent on a patent ductus** to provide flow to the **descending aorta**. Symptoms develop when the aortic ampulla of the ductus closes.
- Less severe obstruction causes no symptoms because blood flow into the descending aorta is not dependent on the ductus.
- Symptoms, including: (may develop before 2 weeks of age)
 1. **poor feeding**
 2. **respiratory distress**
 3. **shock**
- Classically the **femoral pulses** are weaker and delayed compared with the radial pulses.
 - ♥ The blood pressure in the lower extremities is lower than that in the upper extremities.
 - ♥ If cardiac function is poor, these differences may not be apparent until appropriate resuscitation is accomplished.
- There may be no murmur, but an **S₃** is usually present.
- Older children presenting with coarctation of the aorta are usually asymptomatic, although there may be:
 1. a history of **leg discomfort** with exercise
 2. **headache**
 3. **epistaxis**
 4. **Decreased or absent lower extremity pulses**
 5. **hypertension** (upper extremity)
 6. **murmur** may be present
- The murmur of coarctation is typically best heard in the left interscapular area of the back.
- If significant collaterals have developed, continuous murmurs may be heard throughout the chest.
- An abnormal aortic valve is present 50% of the time, causing a **systolic ejection click** and **systolic ejection murmur** of aortic stenosis.

IMAGING STUDIES

- The **ECG** and **chest x-ray** show evidence of **right ventricular hypertrophy** in **infantile coarctation** with marked **cardiomegaly** and **pulmonary edema**.
- In older children, the **ECG** and **chest x-ray** usually show **left ventricular hypertrophy** and a mildly enlarged heart.
- **Rib notching** may also be seen in older children (>8 years old) with large collaterals.
- **Echocardiography** shows:
 1. the site and degree of coarctation
 2. presence of left ventricular hypertrophy
 3. aortic valve morphology and function
 4. associated lesion

TREATMENT

- Management of an infant presenting with cardiac decompensation includes:
 1. **IV infusions of prostaglandin E₁** (chemically opens the ductus arteriosum)
 2. **inotropic agents**
 3. **diuretics**
- **Balloon angioplasty** has been done, especially in critically ill infants
- **surgical repair** of the coarctation is most commonly performed.

