

5TH EDITION

ESSENTIALS of Pediatric Nursing

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Fifth Edition

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As always, I remain motivated by nursing students to seek innovative learning methods for the development of clinical judgment in child health nursing. This book is dedicated to my precious family, my dedicated husband John, my outstanding children Christian and Caitlin, and my amazing granddaughters Sophia and Chloe who continue to inspire me on a daily basis; I am forever grateful for their ongoing faith in and support of me. To Sue Carman, thank you for your ongoing encouragement, dedication, and continual presence, and patience.

—TERRI KYLE

This book continues to be dedicated to the children of the world and the wonderful nurses who care for them. They inspire me to become a better nurse, educator, and person. It is the major impact nurses have on the health of children and their families that drives me to find the best methods to teach clinical judgment in order to provide the very best pediatric nurses that our children deserve. This book is also dedicated to my loving and supportive husband, Chris, without whom I could not have reached this accomplishment; and my four beautiful girls, Grace, Ella, Lily, and Maya, who have allowed me to learn first-hand about growth and development and who truly amaze me each and every day. I would also like to dedicate this to my parents, Lene and Kishor Patel, who always taught me I could do whatever I put my mind to. To Terri Kyle, thank you for this opportunity, your endless support, and your incredible vision.

—SUSAN CARMAN

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ABOUT THE AUTHORS



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Terri Kyle earned a Bachelor of Science in Nursing from the University of North Carolina at Chapel Hill and a Master of Science in Nursing from Emory University in Atlanta, Georgia. Terri received her Doctorate of Nursing Practice in Educational Leadership from American Sentinel

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She has experience in inpatient pediatrics in pediatric and neonatal intensive care units, newborn nursery, specialized pediatric units, and community hospitals. She has worked as a pediatric nurse practitioner in pediatric specialty clinics and primary care. She has been involved in teaching nursing for over 32 years with experience in both undergraduate and graduate nursing education. Terri delights in providing innovative leadership to nursing educators and their students. She is a fellow in the National Association of Pediatric Nurse Practitioners and a member of Sigma Theta Tau International Honor Society of Nursing, the National League for Nursing, and the Society of Pediatric Nurses.



SUSAN CARMAN

Susan Carman earned a Bachelor of Science in Nursing from the University of Wisconsin–Madison and a Master of Science in Nursing and Master in Business Administration from the University of Colorado–Denver. As a pediatric nurse for over 25 years, Susan has had the opportunity to care for children in a variety of diverse settings and in many of the major children's hospitals throughout the United States.

She also has provided volunteer nursing care in a variety of settings including the Dominican Republic and India. She has been involved in teaching nursing for the past 20 years and enjoys watching students transform into competent nurses with strong clinical judgment.

PREFACE

Continuing in the contemporary educational climate, reduced class time is being devoted to specialty courses like pediatric nursing. Therefore, it remains important for nursing educators to focus on key concepts, rather than attempting to cover everything within a specific topic. *Essentials of Pediatric Nursing* was written to direct students to an understanding of critical concepts related to pediatric nursing. Rather than repeating medical-surgical content that the student has already mastered, the text builds upon the student's prior knowledge. The text presents the important differences when caring for children as compared to caring for adults.

The main objectives of the fifth edition of *Essentials of Pediatric Nursing* are to provide student nurses with the foundation needed for high-quality nursing care of children and their families as well as the ability to utilize clinical judgment within various health care environments. The book covers a broad scope of topics, placing emphasis on common issues and pediatric-specific content, including atraumatic care, critical to providing patient and family-centered care. Simpler and broader concepts are mastered first, then students are able to progress to problem-solving in more complex situations.

Reflecting the importance of the nursing process, the steps of assessment, nursing analysis, goal setting with outcome identification, and specific, applicable interventions are provided in the early section of the chapters. This Clinical Judgment and the Nursing Process section provides the student with the general approach needed to care for a child with health alterations specific to that chapter.

Utilizing the nursing process, a concept-based approach provides relevant information in a concise and nonredundant manner. Focusing on conceptual learning provides a time-efficient instructional method for nursing educators and fosters the development of critical thinking in nursing students. Complex critical thinking leads to the development of clinical judgment, which may be applied in various health care environments. This approach is supported by many of the book's features, such as the recurring features, Unfolding Case Studies, Clinical Reasoning Alerts, and Thinking About Development.

ORGANIZATION

Each chapter of *Essentials of Pediatric Nursing* focuses on a different aspect of pediatric nursing care. The book is divided into four units, beginning with general concepts related to pediatric nursing and followed by expected growth and development and specifics related to caring for children. The fourth unit focuses on nursing management of alterations in children's health.

Unit I: Foundations of Pediatric Nursing

Unit I presents the foundational material the nursing student needs to understand how nursing care of the child differs from that of the adult. The unit provides information about general concepts relating to child health. Perspectives on pediatric nursing, the nursing process, and factors influencing child health are key concepts covered in this unit.

Unit II: Health Promotion of the Growing Child and Family

Unit II provides information related to growth and development expectations of the well child from the newborn period through adolescence. Though not exhaustive in nature, this unit provides a broad knowledge base related to normal growth and development that the nurse can draw upon in any situation. Common concerns related to growth and development and child/family education are included in each age-specific chapter.

Unit III: Working With Children and Families

Unit III covers broad concepts that provide the foundation for providing nursing care to children. Rather than reiterating all aspects of nursing care, this unit focuses on specific details needed to provide nursing care for children. The family-centered approach, atraumatic care, communication, and teaching children and families are key concepts in this unit. Additional topics covered in this unit include anticipatory guidance and routine well-child care (including immunizations and safety), health assessment, nursing care of the child in diverse settings, concerns common to special needs children, pediatric variations in nursing procedures, and pain management in children.

Unit IV: Nursing Care of the Child With a Health Disorder

Unit IV focuses on children's responses to health disorders. This unit provides comprehensive coverage of illnesses affecting children. It is arranged according to broad topics of disorders organized with a conceptual and body systems approach and also includes infectious, genetic, and mental health disorders as well as pediatric emergencies. Each chapter follows a similar format to facilitate presentation of the information as well as reduce

repetition. The chapter begins with an overview of variations in pediatric anatomy and physiology, followed by the nursing process for the particular concept. Nursing analysis identifies the patient issue and identified outcomes, and nursing interventions with rationale are included. This approach provides a general framework for addressing alterations in the concept. The nursing process information may then be utilized to develop an individualized nursing care plan or concept map.

RECURRING FEATURES

To provide the student and educator with an exciting and user-friendly text, a number of recurring features have been developed.

Key Terms

Each chapter includes a list of key terms considered to be vital to understanding the content in the chapter. Each key term appears in boldface, with the definition included in the text. Phonetic spellings are provided for terms that may be new or difficult to pronounce.

Learning Objectives

The provision of learning objectives for each chapter helps to guide the student toward prioritizing information for learning. The objectives also provide a method for the student to evaluate understanding of the presented material.

Words of Wisdom

Each chapter opens with inspiring Words of Wisdom (WOW) that set the tone for the chapter. The WOW statements offer students helpful, motivating, or interesting statements that stimulate thinking about children and their families.

Case Studies

Real-life scenarios present relevant child and family information that is intended to improve the student's clinical reasoning skills. Questions throughout the chapter about the scenarios provide an opportunity for the student to critically evaluate the appropriate course of action.

Clinical Reasoning Alert

The Clinical Reasoning Alert promotes critical thinking in the nursing process on information key to clinical reasoning.

Unfolding Patient Stories

Unfolding Patient Stories, written by the National League for Nursing, are an engaging way to begin meaningful conversations in the classroom. These vignettes, which

appear at the end of select chapters, feature patients from Wolters Kluwer's vSim for Nursing | Pediatric (codeveloped by Laerdal Medical) and DocuCare products; however, each Unfolding Patient Story in the book stands alone, not requiring purchase of these products.

Atraumatic Care

These highlights, located throughout the book, provide tips for providing atraumatic care to children in particular situations relating to the topic being discussed.

Take Note!

The *Take Note!* feature draws the student's attention to points of critical emphasis throughout the chapter. This feature is often used to stress vitally important information.

Consider This!

In every chapter, the student is asked to *Consider This!* These first-person narratives engage the student in real-life scenarios experienced by their patients. The personal accounts evoke empathy and help the student to perfect caregiving skills. Each box ends with an opportunity for further contemplation, encouraging the student to think critically about the scenario.

Thinking About Development

The content featured in these boxes will encourage students to think critically about special developmental concerns relating to the topic being discussed.

Healthy People 2030

Throughout the textbook, Healthy People 2030 objectives related to children's health and well-being are outlined in box format. Nursing implications or guidance related to working toward achievement of these objectives is provided. These objectives reflect the Healthy People 2030 guidelines.

Evidence-Based Practice

Throughout the chapters, pertinent questions addressed by current research have been highlighted into Evidence-Based Practice boxes, which discuss recent evidence-based research findings and provide recommendations for nurses.

Teaching Guidelines

Teaching Guidelines, presented in most of the chapters, serve as valuable health education tools. These guidelines raise the student's awareness, provide timely and

accurate information, and are designed to ensure the student's preparation for educating children and their families about various issues.

Drug Guides

The Drug Guide tables summarize information about commonly used medications. The actions, indications, and significant nursing implications presented assist the student in providing optimum care to children and their families.

Common Laboratory and Diagnostic Tests

The Common Laboratory and Diagnostic Tests tables in each chapter of Unit IV provide the student with a general understanding of how a broad range of disorders is diagnosed. Rather than reading the information repeatedly throughout the narrative, the student is then able to refer to the table as needed.

Common Medical Treatments

The Common Medical Treatments tables in each chapter of Unit IV provide the student with a broad awareness of how a common group of disorders is treated either medically or surgically. The table serves as a reference point for common medical treatments.

Comparison Charts

These charts compare two or more disorders or other easily confused concepts. They serve to provide an explanation clarifying the concepts for the student.

Nursing Procedures

Step-by-step nursing procedures provide a clear explanation of pediatric variations to facilitate competent performance.

Concept Mastery Alerts

Concept Mastery Alerts clarify pediatric nursing concepts to improve the reader's understanding of potentially confusing topics as identified by Misconception Alerts in Lippincott's Adaptive Learning Powered by PrepU. Data from thousands of actual students using this program in courses across the United States identified common misconceptions for the authors to clarify in this new feature.

Dosage Calculation Box

This box provides a dosage calculation example in each of the alteration/disorder chapters. Reiteration of the significance of accurate dosage calculation assists the student with mastery of this critical concept.

Key Concepts

At the end of each chapter, Key Concepts provide a quick review of essential chapter elements. These bulleted lists help the student focus on the important aspects of the chapter.

Tables, Boxes, Illustrations, and Photographs

Tables and boxes are included throughout the chapters to summarize key content areas. Beautiful illustrations and photographs help the student to visualize the content. These features allow the student to quickly and easily access information.

References

References that were used in the development of the text are provided at the end of each chapter. The listings allow the student to further pursue topics of interest.

Developing Clinical Judgment

This section located at the end of each chapter assists the student with the development of clinical judgment through:

- **Practicing for NCLEX**—these NCLEX-RN style questions test the student's ability to utilize critical thinking in the application of the nursing process to chapter material. The questions are styled similarly to the national licensing examination. Next-Gen NCLEX-RN style questions are also included in most chapters.
- **Dosage Calculation Questions**—these problems test the student's ability to accurately determine medication dosages particular to children.
- **Critical Thinking Exercises**—these exercises serve to stimulate the student to incorporate the current material with previously learned concepts and reach a satisfactory conclusion. The exercises encourage students to think critically, problem solve, and consider their own perspective on given topics.
- **Study Activities**—these activities promote student participation in the learning process. This section encourages increased interaction/learning via clinical, online, and community activities.
- **Answers**—answers to the Developing Clinical Judgment questions are provided to instructors on [thePoint®](#).

A NOTE ABOUT THE LANGUAGE USED IN THIS BOOK

Wolters Kluwer recognizes that people have a diverse range of identities, and we are committed to using inclusive and nonbiased language in our content. In line with

the principles of nursing, we strive not to define people by their diagnoses, but to recognize their personhood first and foremost, using as much as possible the language diverse groups use to define themselves, and including only information that is relevant to nursing care.

We strive to better address the unique perspectives, complex challenges, and lived experiences of diverse populations traditionally underrepresented in health literature. When describing or referencing populations discussed in research studies, we will adhere to the identities presented in those studies to maintain fidelity to the evidence presented by the study investigators. We follow best practices of language set forth by *the Publication Manual of the American Psychological Association, 7th edition*, but acknowledge that language evolves rapidly, and we will update the language used in future editions of this book as necessary.

INSTRUCTOR'S RESOURCES

Tools to assist instructors with teaching the course are available through **thePoint®**, upon adoption of this text.

- An **E-Book** on **thePoint®** gives you access to the book's full text and images online.
- A **Test Generator** lets you put together exclusive new tests from a bank containing **hundreds of questions** to help you in assessing your students' understanding of the material. Test questions link to chapter learning objectives.
- **PowerPoint presentations with Guided Lecture Notes** provide an easy way for you to integrate the textbook with your students' classroom experience, either via slide shows or handouts. Multiple choice and true/false questions are integrated into the presentations to promote class participation and allow you to use i-clicker technology.
- An **Image Bank** lets you use the photographs and illustrations from this textbook in your PowerPoint slides or as you see fit in your course.
- An **AACN Essentials map** relates the textbook content to the current AACN Essentials.
- A sample **Syllabus** provides guidance for structuring your pediatric nursing course.
- **Journal Articles**, updated for the new edition, offer access to current research available in Wolters Kluwer journals.

Contact your sales representative or check out LWW.com/Nursing for more details and ordering information.

Lippincott CoursePoint+

This text is also available for sale in the **Lippincott CoursePoint+** version.

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- Powerful tools, including varying levels of case studies, interactive learning activities, and adaptive learning powered by PrepU, help students learn the critical thinking and clinical judgment skills to help them become practice-ready nurses.
- Preparation for Practice tools improve student competence, confidence, and success in transitioning to practice.
 - vSim® for Nursing: Codeveloped by Laerdal Medical and Wolters Kluwer, vSim® for Nursing simulates real nursing scenarios and allows students to interact with virtual patients in a safe, online environment.
 - Lippincott® Advisor for Education: With over 8500 entries covering the latest evidence-based content and drug information, Lippincott® Advisor for Education provides students with the most up-to-date information possible, while giving them valuable experience with the same point-of-care content they will encounter in practice.
- Unparalleled reporting provides in-depth dashboards with several data points to track student progress and help identify strengths and weaknesses.
- Unmatched support includes training coaches, product trainers, and nursing education consultants to help educators and students implement CoursePoint with ease.

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WOW

WORDS OF WISDOM

The heart of the matter is healing the child's heart so they can embrace life to its fullest.

19



Nursing Care of the Child With an Alteration in Perfusion/Cardiovascular Disorder

LEARNING OBJECTIVES

Upon completion of the chapter, you will be able to:

1. Compare anatomy and physiology of the cardiovascular system in infants and children with that of adults.
2. Describe nursing care related to common laboratory and diagnostic tests used in the medical diagnosis of pediatric cardiovascular conditions.
3. Distinguish cardiovascular disorders common in infants, children, and adolescents.
4. Identify appropriate nursing assessments and interventions related to medications and treatments for pediatric cardiovascular disorders.
5. Develop an individualized nursing care plan or concept map for the child with a cardiovascular disorder.
6. Describe the psychosocial impact of chronic cardiovascular disorders on children.
7. Devise a nutrition plan for the child with cardiovascular disease.
8. Develop child and family teaching plans for the child with a cardiovascular disorder.

KEY TERMS

arrhythmia

cardiomegaly (kahr'dē-ō-meg'ă-lē)

clubbing

echocardiography

electrocardiogram

heart failure

orthotopic (or'thō-tō'pik)

polycythemia (pol'ē-sī-thē'mē-ă)

Logan Bernstein, 6 weeks old, is brought to the clinic by his parent. He presents with poor feeding. His parent states, "Logan falls asleep while feeding, and he's always sweaty during feedings."

INTRODUCTION

Perfusion refers to the mechanisms that facilitate blood through tissue. Nurses may encounter alterations in perfusion in children and should be familiar with various cardiovascular disorders that children experience. Alterations in perfusion, or cardiovascular disease, are a significant cause of chronic illness and death in children. Typically, cardiovascular disorders in children are divided into two major categories—congenital heart defects (CHDs) and acquired heart disease.

CHD is defined as structural anomalies that are present at birth, although they are often not diagnosed until later in life. About 40,000 babies are born annually with CHD, accounting for the largest percentage of all birth defects (American Heart Association [AHA], 2022). CHD may result from a genetic abnormality or be associated with a genetic syndrome. About 40% to 50% of children with Down syndrome have a CHD (Nees & Chung, 2020). Many CHDs result in **heart failure** (inability of the heart to pump blood sufficiently) and chronic cyanosis, leading to failure to thrive.

Acquired heart disease includes disorders that occur after birth. These disorders develop from a wide range of causes, or they can occur as a complication or long-term effect of CHD.

The diagnosis of a cardiovascular disorder in any person can be extremely frightening and overwhelming. Early on, children learn that the heart is necessary for life, so knowing that there is a heart problem can promote feelings of dread. These feelings are compounded by the child's age, the view of the child as being vulnerable and defenseless, and the stressors associated with the disorder itself. The child and parents need much support and reassurance (Gaskin & Kennedy, 2019).

Nurses need to have a sound knowledge base about cardiovascular conditions affecting children so that they can provide appropriate assessment, intervention, guidance, and support to the child and family. Cardiovascular disorders require acute interventions that often have long-term implications for the child's health and growth and development. Due to the potentially overwhelming and devastating effects that cardiovascular disorders can have on children and their families, nurses need to be skilled in assessment and interventions in this area and able to provide support throughout the course of the illness and beyond.

VARIATIONS IN PEDIATRIC ANATOMY AND PHYSIOLOGY

The cardiovascular system undergoes numerous changes at birth. Structures that were vital to the fetus are no longer needed. Circulation via the umbilical arteries and

vein is replaced with the child's own closed independent circulation. Changes in the size of the heart, pulse rate, and blood pressure (BP) also occur.

Circulatory Changes From Gestation to Birth

The fetal heart rate is present on about postconceptual day 17. The four chambers of the heart and arteries are formed during gestational weeks 2 through 8, with maturation of the structures occurring throughout the remainder of gestation. During fetal development, oxygenation of the fetus occurs via the placenta; the lungs, although perfused, do not perform oxygenation and ventilation. The foramen ovale, an opening between the atria, allows blood flow from the right to the left atrium. The ductus arteriosus allows blood flow between the pulmonary artery and the aorta, shunting blood away from the pulmonary circulation (Cunningham et al., 2022). Figure 19.1 illustrates fetal circulation.

With the newborn's first breath, several changes occur in the cardiopulmonary system that enable the newborn to make the transition from fetal circulation to normal circulation. As the newborn breathes for the first time, the lungs inflate, reducing pulmonary vascular resistance to blood flow. As a result, pulmonary artery pressure drops. Subsequently, pressure in the right atrium decreases. Blood flow to the left side of the heart increases the pressure in the left atrium. This change in pressure leads to closure of the foramen ovale. The drop in pressure of the pulmonary artery promotes closure of the ductus arteriosus, which is located between the aorta and the pulmonary artery. The ductus venosus, located between the left umbilical vein and the inferior vena cava, closes because of a lack of blood flow and vasoconstriction. The closed ductus arteriosus and ductus venosus eventually become ligaments. With the lack of blood flow to the umbilical arteries and vein, these structures atrophy (Cunningham et al., 2022).

Structural and Functional Differences

The structure and function of the infant's and child's cardiovascular system differ from those of adults, depending on age. In infants and children younger than 7 years, the heart lies more horizontally, resulting in the apex lying higher in the chest, below the fourth intercostal space. As the lungs grow over time, the heart is displaced downward. Between ages 1 and 6 years, the heart is four times the birth size. Between 6 and 12 years of age, the child's heart is 10 times the size it was at birth. However, the heart is smaller proportionally at this time than at any other stage in life. During the school-age years, the heart

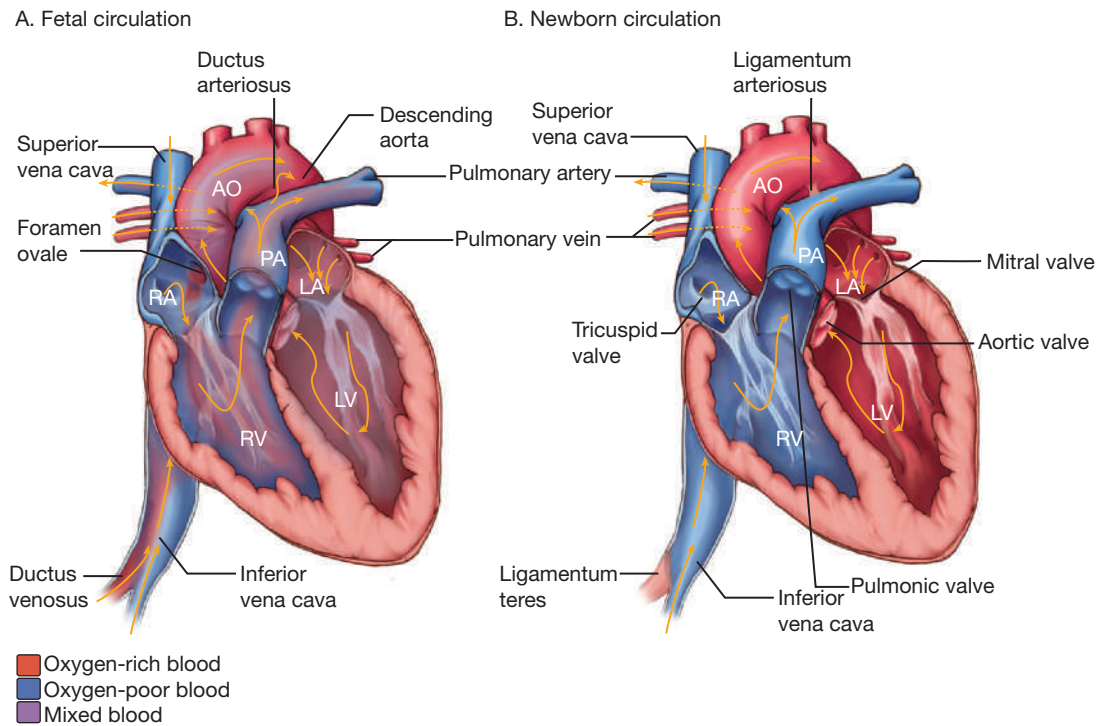


FIGURE 19.1 Fetal and newborn circulation.

grows more vertically within the thoracic cavity. During adolescence, the heart continues to grow in relation to the adolescent's rapid growth.

At birth, the ventricle walls are similar in thickness, but with time the left ventricular wall thickens. The immature myocytes of the infant's heart are thinner and less compliant than those of the adult. Right ventricular function dominates at birth, and over the first few months of life, left ventricular function becomes dominant. The infant's heart at rest exhibits a greater resting tension than the adult's, so volume loading or increased stretch may actually lead to decreased cardiac output. The infant's sarcoplasmic reticulum is less well organized than the adult's, making the infant dependent on serum calcium for contraction. Inotropic response to calcium in the actin and myosin (contractile proteins) increases with age.

The normal heart rate is higher in infancy than in adulthood, limiting the infant's ability to increase cardiac output by increasing the heart rate. The heart's efficiency increases as the child ages and the heart rate drops over time. The normal infant heart rate ranges from 90 to 160 beats per minute (bpm), the toddler's or preschooler's is 80 to 115 bpm, and the school-age child's and the adolescent's ranges from 60 to 100 bpm. Innocent murmurs and physiologic splitting of heart sounds may be noted in infancy or childhood. These findings are related to the change in the size of the heart in relation to the thoracic cavity. The infant's and child's blood vessels widen and increase in length over time. The average infant's BP is about 80/55 mm Hg; BP is usually lower in the younger infant and

can be slightly higher in the older infant. The BP increases over time to the adult level. The toddler or preschooler's BP averages 90 to 110/55 to 75 mm Hg, the school-age child's 100 to 120/60 to 75 mm Hg, and the adolescent's 100 to 120/70 to 80 mm Hg (Kleinman et al., 2021).

COMMON MEDICAL TREATMENTS

A variety of medications as well as other medical treatments and surgical procedures are used to treat cardiovascular problems in children. Most of these treatments will require a health care provider's order when the child is in the hospital. The most common treatments and medications are listed in Common Medical Treatments 19.1 and Drug Guide 19.1. The nurse caring for the child with a cardiovascular disorder should be familiar with what the procedures and medications are and how they work as well as common nursing implications related to use of these modalities.

TAKE NOTE!

Give digoxin at regular intervals, every 12 hours, such as at 8 a.m. and 8 p.m., 1 hour before or 2 hours after a feeding. If a digoxin dose is missed, give the dose as soon as it is realized the dose was missed. If it is close to the next dose's time, hold the missed dose. Monitor potassium levels, as a decrease enhances the effects of digitalis, causing toxicity (UpToDate, Inc., 2024).

COMMON MEDICAL TREATMENTS 19.1

Treatment	Explanation	Indications	Nursing Implications
Oxygen	Supplemented via mask, nasal cannula, hood, tent, or endotracheal/nasotracheal tube	Hypoxemia, respiratory distress, heart failure	Monitor response via work of breathing and pulse oximetry.
Chest physiotherapy (CPT) and postural drainage	Promotes mucus clearance by mobilizing secretions with the assistance of percussion or vibration accompanied by postural drainage (refer to Chapter 18 for additional information related to CPT and postural drainage)	Mobilization of secretions, particularly in postoperative period or with heart failure	May be performed by respiratory therapist in some institutions, by nurses in others; in either case, nurses must be familiar with the technique and able to educate families on its use.
Chest tube	Drainage tube is inserted into the pleural cavity to facilitate removal of air or fluid and allow full lung expansion.	After open heart surgery, pneumothorax	If tube becomes dislodged from container, the chest tube must be clamped immediately to avoid further air entry into the chest cavity. Alternatively, the end may be immediately placed into a container of sterile water or saline to create a water seal.
Pacing	External wiring connected to a small generator used to electrophysiologically correct cardiac arrhythmias or heart block (temporary). Permanent pacing achieved with an implantable internal pacemaker.	Bradycardias, heart block, cardiomyopathy, sinoatrial or atrioventricular node malfunction	Provide close observation of the child, pacing unit, and ECG. Maintain asepsis at pacing lead insertion site. Explain to the child and family that the permanent pacemaker may be felt under the skin. Advise against participation in contact sports. ^a

ECG, electrocardiogram

^aChildren's Hospital of Wisconsin. (2024). *Living with a pediatric pacemaker*. <https://childrenswi.org/medical-care/herma-heart/conditions/living-with-a-pacemaker>

DRUG GUIDE 19.1

Medication	Actions/Indications	Nursing Implications
Alprostadil (prostaglandin)	Direct vasodilation of the ductus arteriosus smooth muscle Indicated for temporary maintenance of ductus arteriosus patency in infants with ductal-dependent congenital heart defects	<ul style="list-style-type: none"> • Apnea occurs in 10%–20% of neonates within first hour of infusion. • Monitor arterial BP, respiratory rate, heart rate, ECG, temperature, and pO₂; watch for abdominal distention. • Fresh IV solution required every 24 hours. • Reposition catheter if facial or arm flushing occurs. • Use with caution in neonate with bleeding tendency. • Contraindicated in respiratory distress syndrome or persistent fetal circulation
Digoxin (cardiac glycoside, antiarrhythmic agent)	Increases contractility of the heart muscle by decreasing conduction and increasing force Used for heart failure, atrial fibrillation, atrial flutter, supraventricular tachycardia	<ul style="list-style-type: none"> • Prior to administering each dose, count apical pulse for one full minute, noting rate, rhythm, and quality. Withhold if apical pulse is <60 in an adolescent, <90 in an infant. • Avoid giving oral form with meals, as altered absorption may occur. • Monitor serum digoxin levels (therapeutic range: 0.8–2 ng/mL). • Note signs of toxicity: nausea, vomiting, diarrhea, lethargy, and bradycardia. • Ginseng, hawthorn, and licorice intake increase risk of drug toxicity. • Note contraindications (ventricular fibrillation and hypersensitivity to digitalis). • Avoid rapid IV administration, as this may lead to systemic and coronary artery vasoconstriction.

(continued)

DRUG GUIDE 19.1 (continued)

Medication	Actions/Indications	Nursing Implications
Furosemide (loop diuretic)	Inhibits resorption of sodium and chloride Used to manage edema associated with heart failure, and hypertension in combination with antihypertensives	<ul style="list-style-type: none"> Administer with food or milk to decrease GI upset. Monitor BP, kidney function, electrolytes (particularly potassium), and hearing. May cause photosensitivity
Heparin (anticoagulant)	Interferes with conversion of prothrombin to thrombin, preventing clot formation Indicated for the prophylaxis and treatment of thromboembolic disorders, especially after cardiac surgery	<ul style="list-style-type: none"> Administer SQ, not IM. Dose is adjusted according to coagulation test results. Monitor for signs of bleeding, platelet counts. Ensure that the antidote, protamine sulfate, is available. Do not administer with uncontrolled bleeding or if subacute bacterial endocarditis is suspected.
Indomethacin (nonsteroidal antiinflammatory agent)	Inhibits prostaglandin synthesis in order to close patent ductus arteriosus	<ul style="list-style-type: none"> Monitor heart rate, BP, ECG, and urine output; monitor for murmur. Monitor serum sodium, glucose, platelet count, BUN, creatinine, potassium, and liver enzymes. May mask signs of infection Note development of edema.
Spironolactone (potassium-sparing diuretic)	Competes with aldosterone to result in increased water and sodium excretion (saves potassium) Used to manage edema due to heart failure and for treatment of hypertension	<ul style="list-style-type: none"> Administer with food. Monitor serum potassium, sodium, and kidney function. May cause drowsiness, headache, and arrhythmia May cause false elevations in digitalis level Teach children to avoid high-potassium diets, salt substitutes, and natural licorice. Contraindicated in hyperkalemia, kidney failure, and anuria
Antibiotics		
Penicillin G benzathine Penicillin V potassium	Inhibits bacterial wall synthesis in susceptible organisms. Indicated in mild to moderate infections, for prophylaxis of endocarditis and rheumatic fever	<ul style="list-style-type: none"> Contraindications include hypersensitivity to penicillins. Report hypersensitivity reactions (chills, fever, wheezing, pruritus, anaphylaxis) immediately. PCN-G: administer IM. Pen-VK: administer orally on empty stomach 1 hour before or 2 hours after a meal.
Erythromycin (macrolide)	Inhibits ribonucleic acid transcription in susceptible organisms. Used in children with penicillin allergy, mild to moderate infections, or endocarditis, and for rheumatic fever prophylaxis	<ul style="list-style-type: none"> Contraindicated in preexisting liver disease IV administration may result in CV abnormalities. Abdominal distress common with oral use. Fever, dizziness, and rash may occur.
Antihypertensive Drugs		
Angiotensin-converting enzyme (ACE) inhibitors (captopril, enalapril)	Competitive inhibition of ACE for management of hypertension Heart failure management in conjunction with digitalis and diuretics	<ul style="list-style-type: none"> Monitor BP, kidney function, WBC count, and serum potassium. Discontinue if angioedema occurs. Captopril: administer orally on empty stomach 1 hour before or 2 hours after meals. Enalapril: may administer orally without regard to food
Beta-adrenergic blockers (propranolol, atenolol, sotalol)	Competitively block response to beta-adrenergic stimulation, decreasing heart rate, and force of contraction. Used for management of hypertension, arrhythmias, and prevention of myocardial infarction	<ul style="list-style-type: none"> Monitor ECG and BP. Propranolol: administer with food. Atenolol, sotalol: administer without regard to food. Do not stop drug abruptly. May result in bradycardia, dizziness, nausea and vomiting, dyspnea, and hypoglycemia (propranolol) Contraindications: heart block, uncompensated heart failure, cardiogenic shock, asthma, or hypersensitivity
Hydralazine (vasodilator)	Direct vasodilation of arterioles to manage moderate to severe hypertension, heart failure	<ul style="list-style-type: none"> Monitor heart rate and BP. Closely monitor BP with IV use. Administer oral dose with food. May cause palpitations, flushing, tachycardia, dizziness, nausea, and vomiting Notify health care provider or nurse practitioner if flu-like symptoms occur. Contraindicated in rheumatic valvular disease

BP, blood pressure; BUN, blood urea nitrogen; CV, cardiovascular; ECG, electrocardiogram; GI, gastrointestinal; IM, intramuscularly; IV, intravenous; pO₂, partial pressure of oxygen; SQ, subcutaneously; WBC, white blood cell.

Source: UpToDate, Inc. (2024). *UpToDate@ Lexidrug™* (Version 8.2.0) [Mobile app]. Wolters Kluwer. <https://apps.apple.com/us/app/lexicomp/id313401238>

COMMON LABORATORY AND DIAGNOSTIC TESTS 19.1

Test	Explanation	Indications	Nursing Implications
Arteriogram (angiogram: visualization of arteries or veins)	Radiopaque contrast solution is injected through a catheter and into the circulation. Radiographs are then taken to visualize the structure of the heart and blood vessels.	To observe blood flow to parts of body and detect lesions; to confirm a diagnosis Catheters can be used to remove plaques.	<ul style="list-style-type: none"> • Make sure the parent signs a consent form. • Administer premedication as ordered. • Obtain child's weight to determine amount of dye needed. • Keep the child NPO before the procedure according to institutional protocol. • After the procedure, maintain the child on bed rest. • Observe the puncture site for bleeding. • Monitor vital signs frequently and check the pulse distal to the site.
Ambulatory electrocardiographic monitoring (Holter)	Monitoring of the heart's electrical patterns for 24 hours using a portable compact unit	To identify and quantify arrhythmias in a 24-hour period during normal daily activities	<ul style="list-style-type: none"> • Instruct the child and parent to push the "event button" whenever chest pain, syncope, or palpitations occur. • Normal daily activities should be carried out during the testing period. • Having the child wear a snug undershirt over the leads helps to keep them in place.
Chest radiograph	A radiographic film of the chest area; will determine size of the heart and its chambers and pulmonary blood flow	Serves as a baseline for comparison with films taken after surgery; used to identify abnormalities of the lungs, heart, and other structures in the chest	<ul style="list-style-type: none"> • Instruct child not to wear jewelry or any metal around neck or on the hospital gown. • Explain to the child and family that no pain or discomfort should result. • If a portable radiograph at bedside is done, remove electrodes temporarily.
Echocardiogram	Noninvasive ultrasound procedure used to assess heart wall thickness, size of heart chambers, motion of valves and septa, and relationship of great vessels to other cardiac structures	Specific diagnosis of structural defects; determines hemodynamics and detects valvular defects	<ul style="list-style-type: none"> • Assure the child that the echo does not hurt. • Instruct the child about ECG lead placement and use of gel on the scope's wand during the procedure. • Encourage the child to lie still throughout the test.
Electrocardiogram (ECG)	A graphic record produced by an electrocardiograph (device used to record the electrical activity of the myocardium to detect transmission of the cardiac impulse through the conductive tissues of the muscle) Facilitates evaluation of the heart rate, rhythm, conduction, and musculature.	To detect heart rhythm and chamber overload; also serves as a baseline for measuring postoperative complications.	<ul style="list-style-type: none"> • Assure the child that monitoring is a painless procedure. • Place electrodes in the appropriate location. • The child must lie still during the ECG recording period (usually about 5 minutes). • Wipe electrode paste or jelly off after procedure.
Exercise stress test	Monitoring of heart rate, blood pressure, ECG, and oxygen consumption at rest and during exercise	Quantifies exercise tolerance; can be used to provoke symptoms or arrhythmias	<ul style="list-style-type: none"> • Child should be NPO for 4 hours prior to test. • Obtain baseline ECG and vital signs. • Instruct child to verbalize symptoms during testing. • Usually takes about 45 minutes.
Hemoglobin (Hgb) and hematocrit (Hct)	Measures the total amount of hemoglobin in the blood and indirectly measures the red blood cell number and volume.	To detect anemia or polycythemia (may occur with CHD resulting in cyanosis)	<ul style="list-style-type: none"> • False elevations occur with dehydration. • May be obtained quickly via capillary puncture • Normal values vary with age.
Partial pressure of oxygen (pO ₂)	Measures the amount of oxygen in the blood.	To determine the presence and degree of hypoxia	<ul style="list-style-type: none"> • Most accurate result is with arterial specimen (venous and capillary specimens demonstrate lower levels). • Observe child for cyanosis. • Supplement with oxygen per protocol.
Pulse oximetry screening	Noninvasive method of measuring oxygen saturation in the blood	To detect critical congenital heart disease in the newborn	<ul style="list-style-type: none"> • Take measurements in the right hand and in either foot. Apply the probe correctly and securely, being sure to minimize movement and ambient light interference.

CHD, congenital heart defect; NPO, nothing by mouth

Data from Children's Heart Institute. (2023). *Heart tests? When do you need them?* <https://www.childrensheartinstitute.org/health-library/health-wise/?DOCHWID=aba5713>; Corbett, J. A., & Banks, A. D. (2019). *Laboratory tests and diagnostic procedures with nursing diagnoses* (9th ed.).

Pearson Education Inc; Oster, M. (2023). Newborn screening for critical congenital heart disease using pulse oximetry. *UpToDate*. Retrieved March 31,

2023, from <https://www.uptodate.com/contents/newborn-screening-for-critical-congenital-heart-disease-using-pulse-oximetry>

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CARDIAC CATHETERIZATION

Cardiac catheterization is the definitive study for infants and children with cardiac disease. It has become almost a routine diagnostic procedure and may be performed on an outpatient basis. However, it is highly invasive and not without risks, especially in sick infants and children. Indications for cardiac catheterization include:

- Cardiovascular disease, causing cyanosis in infants; these infants need to be catheterized as soon as they are in a reasonably stable condition.
- Severe heart failure or progressive problems such as pulmonary edema
- Questionable anatomic or physiologic abnormalities
- Planned cardiac surgery
- Progressive monitoring related to pulmonary hypertension
- Periodic assessment after repair of a cardiac defect
- Therapeutic interventions such as septostomy or balloon valvotomy

Cardiac catheterization may be categorized as diagnostic or interventional. The type of catheterization performed varies based on the individual needs of the child. The procedure lasts from 2 to 5 hours (UPMC, 2024).

In cardiac catheterization, a radiopaque catheter is inserted into a blood vessel and is then guided through the vessel to the heart with the aid of fluoroscopy. For a right-sided catheterization, the catheter is threaded to the right atrium via a major vein such as the femoral vein. With a left-sided catheterization, the catheter is threaded to the aorta and heart via an artery. Once the tip of the catheter is in the heart, contrast material is injected via the catheter, and radiographic images are taken.

While the catheter is in the heart, several procedures can be performed. The BP, changes in cardiac output or stroke volume, and oxygen saturation in each heart chamber and major blood vessels are recorded. With the injection of contrast material, information is revealed about the heart anatomy, ventricular wall motion and ejection fraction, intracardiac pressures and hemodynamic parameters, cardiac valve function, and structural abnormalities. The movement of the contrast material is filmed so that the details of the cardiac procedure are recorded. Samples of heart tissue to evaluate for infection, muscular dysfunction, or rejection after a transplant may also be obtained (UPMC, 2024).

Clinical Judgment and the Nursing Process

Care of the child with a cardiovascular disorder includes all steps of the nursing process: assessment, nursing analysis, planning, interventions, and evaluation. There are a number of general concepts related to the nursing process that may be applied to any child

with a cardiovascular disorder. The nurse should be knowledgeable about the procedures, treatments, and medications as well as familiar with the nursing implications related to these interventions. With an understanding of these concepts, the nurse can individualize the care based on the child's and family's needs.

Assessment

When assessing a child with a cardiovascular disorder, expect to obtain a health history, perform a physical examination, and prepare the child for laboratory and diagnostic testing.

Health History

The health history consists of a history of the present illness, past medical history, and family history. Depending on their age, the child should be included in the health history interview; the child's age will determine the degree of involvement and the terminology used. Table 19.1 gives examples of typical questions that can be used when obtaining the child's health history.

HISTORY OF PRESENT ILLNESS

Elicit the history of the present illness, which addresses when the symptoms started and how they have progressed. Inquire about any treatments and medications used at home. Ask parents about history of orthopnea, dyspnea, easy fatigability, growth delays, squatting, edema, dizziness, and/or frequent occurrences of pneumonia, which can be significant signs of pediatric heart disease. The history of present illness may reveal a history of poor feeding, including fatigue, lethargy, and/or vomiting, or failure to thrive, even with adequate caloric intake. The parents may report diaphoresis, which is often seen in early heart failure. The parent or caregiver may also report delays in gross motor development, cyanosis (possibly reported by the parents as more of a gray color than blue), and tachypnea (indicative of heart failure).

PAST MEDICAL HISTORY

The past medical history includes information about the child and the birthing parent's pregnancy history. Assess the child's past medical history for:

- Problems occurring after birth (history of the child's condition after birth may reveal evidence of an associated congenital malformation or other disorder.)
- Frequent infections
- Chromosomal abnormalities
- Prematurity
- Autoimmune disorders
- Use of medications, such as corticosteroids

Assess the birthing parent's pregnancy, labor, and delivery history. Be sure to include information about the status of the neonate at birth. Also inquire about the birthing parent's use of medications, including illicit

TABLE 19.1 • Examples of Questions for Obtaining a Child's Health History

Questions	Provides Information About
<ul style="list-style-type: none"> • What types and amounts (dosages) of medications has the child received? What were they used for? • Who prescribed them? • Were they effective? Did the child experience any adverse effects? • To whom does the child go for medical evaluation? How often? Were the visits for regular health check-ups or for situational problems? Were there previous hospitalizations? What for? • Has the child experienced any growth delay? Does the child have any problems with activity and coordination? • Does the child's skin color change when crying? If so, what color do you see? • Does the child stop frequently during play to sit or squat? • Does the child have feeding difficulty? Does the child tire easily or sleep excessively? • Does the child frequently develop strep throat? 	<ul style="list-style-type: none"> • Possible underlying conditions that may be related to the child's current status • Other healthcare personnel involved in the child's care as well as the parents' health care beliefs and patterns • How the medications may be affecting the child's health • The child's health status and the parents' healthcare knowledge, practices, and beliefs • Problems that may result from impaired cardiac output, adequacy of tissue oxygenation, and concomitant disorders associated with heart disease • Effectiveness of tissue oxygenation. A blue or gray skin color may be due to cyanosis. • The child's exercise tolerance and tissue oxygenation • The child's energy expenditure, ability to tolerate activity, and tissue oxygenation • The child's risk for developing rheumatic fever and heart disease

Data from Hueckel, R. M. (2019). Pediatric patient with congenital heart disease. *Journal for Nurse Practitioners*, 15(1), 118–124. <https://doi.org/10.1016/j.nurpra.2018.10.017>

or over-the-counter drugs and alcohol; exposure to radiation; presence of hypertension; and viral illnesses such as coxsackievirus, cytomegalovirus, influenza, mumps, or rubella. A history of significant problems related to labor and delivery is also important: stress or asphyxia at birth may be related to cardiac dysfunction and pulmonary hypertension in the newborn.

Assess for additional risk factors such as:

- Family history of heart disease or CHD (investigate the history further if heart disease occurred in a first-degree relative)
- Hyperlipidemia
- Diabetes mellitus
- Obesity
- Inactivity
- Stress
- High-cholesterol diet

PHYSICAL EXAMINATION

Physical examination of the child with a cardiovascular condition consists of inspection, palpation, and auscultation. In addition, obtain the child's vital signs and measure the child's height and weight. Plot this information on a standard growth chart to evaluate nutritional status and growth. If the child is younger than 3 years, measure and plot the head circumference also.

INSPECTION

Assess the child's overall appearance. Inspect the color of the skin, noting cyanosis. Inspect the skin for edema. In infants, peripheral edema occurs first in the face, then the presacral region, and then the extremities. Edema of the lower extremities is characteristic of right ventricular heart failure in older children.



CLINICAL REASONING ALERT!

Suspect CHD in the cyanotic newborn who does not improve with oxygen administration (Weiner et al., 2021).

Inspect the fingers and toes for clubbing. Clubbing (which usually does not appear until after 1 year of age) implies chronic hypoxia due to severe CHD. The first sign of **clubbing** is softening of the nail beds, followed by rounding of the fingernails, followed by shininess and thickening of the nail ends (see Fig. 18.5 in Chapter 18). Obtain the child's temperature; fever would suggest infection. Assess respirations, including rate, rhythm, and effort. Note location and severity of retractions if present. Inspect the chest configuration, noting any prominence of the precordial chest wall, which is often seen in infants and children with **cardiomegaly** (abnormal heart enlargement). Note visible pulsations, which may indicate increased heart activity. Also inspect the neck veins for engorgement or abnormal pulsations. Note abdominal distention.



CLINICAL REASONING ALERT!

Children with cardiac conditions resulting in cyanosis will often have baseline oxygen saturations that are relatively low because of the mixing of oxygenated with deoxygenated blood.

PALPATION

Palpate the right and left radial or brachial pulse to assess cardiac rate and rhythm. Throughout infancy and childhood, the rate may vary. Palpate the femoral pulse; it should be readily palpable and equal in amplitude

and strength to the brachial or radial pulse. A femoral pulse that is weak or absent in comparison to the brachial pulse is associated with coarctation of the aorta. Significant variations in pulse occur with activity, so the most accurate heart rate may be determined during sleep. In older children, exercise and emotional factors may influence the heart rate. A bounding pulse is characteristic of patent ductus arteriosus (PDA) or aortic regurgitation. Narrow or thready pulses may occur in children with heart failure or severe aortic stenosis (Driscoll, 2022). Note tachycardia, bradycardia, rhythm irregularities, diminished peripheral pulses, or thready pulse. Palpate the child's abdomen for hepatomegaly, a sign of right-sided heart failure in the infant and child.

AUSCULTATION

Auscultate the apical pulse for a full minute to determine heart rate and rhythm. Note irregularities in rhythm, tachycardia, or bradycardia. Auscultate the heart for murmurs. Many children have functional or innocent murmurs, but all murmurs must be evaluated on the basis of the following characteristics:

- Location
- Relation to the heart cycle and duration
- Intensity: grade I, soft and hard to hear; grade II, soft and easily heard; grade III, loud without thrill; grade IV, loud with a precordial thrill; grade V, loud with a precordial thrill, audible with a stethoscope partially off chest; grade VI, very loud, audible with a stethoscope or with the naked ear
- Quality: harsh, musical, or rough; high, medium, or low pitch
- Variation with position (sitting, lying, standing) (Driscoll, 2022)

Auscultate for the character of heart sounds. Note distinct, muffled, or distant heart sounds. Abnormal splitting or intensifying of S_2 sounds occurs in children with major heart problems. Ejection clicks, which are high pitched, are related to problems with dilated vessels and/or valve abnormalities. Heard throughout systole, they can be early, moderate, or late. Clicks on the upper left sternal border are related to the pulmonary area. Aortic clicks are best heard at the apex and can be mitral or aortic in origin. A mild to late ejection click at the apex is typical of a mitral valve prolapse. The S_3 heart sound may be heard in children, diminishing when moving from supine to upright, and a pathologic S_3 may occur with poor cardiac function. The S_4 heart sound is not normally audible and is associated with cardiomyopathy or diastolic dysfunction (Jone et al., 2022).

Auscultate the BP in the upper extremities and lower extremities, and compare the findings; there should be no major differences between the upper and lower extremities. Determine the pulse pressure by subtracting the diastolic pressure from the systolic pressure. The pulse pressure is less than 50 mm Hg, or

less than half the systolic pressure. A widened pulse pressure, which is usually accompanied by a bounding pulse, is associated with PDA, aortic insufficiency, fever, anemia, or complete heart block. A narrowed pulse pressure is associated with aortic stenosis. Note hypotension or hypertension.

TAKE NOTE!

Alert children and parents if a heart murmur is detected, even if it is benign.

Laboratory and Diagnostic Testing

Common Laboratory and Diagnostic Tests 19.1 explains the laboratory and diagnostic tests most commonly used when considering cardiovascular disorders in children. The tests can assist the health care provider in diagnosing the disorder or can be used as guidelines in determining ongoing treatment. Laboratory or non-nursing personnel obtain some of the tests, while the nurse might obtain others. In either instance, the nurse should be familiar with how the tests are obtained, what they are used for, and normal versus abnormal results. This knowledge will also be necessary when providing child and family education related to the testing.

Remember Logan, the 6-week-old with poor feeding? What additional health history and physical examination assessment information should the nurse obtain?

Nursing Analysis

After recognizing and analyzing cues from a thorough assessment, the nurse might identify several patient problems, including:

- Decreased cardiac output
- Excess fluid volume
- Activity intolerance
- Imbalanced nutrition, less than body requirements
- Risk of delayed development
- Pain
- Interrupted family processes
- Deficient knowledge

After completing Logan's assessment, the nurse noted the following: poor weight gain, tachypnea with occasional nasal flaring, crackles heard on auscultation, and edema noted in the face, presacral area, and extremities. Based on these assessment findings, what would your top three concerns be for Logan?

The foregoing patient issues provide suggestions for nursing care planning or concept mapping. Suggested interventions with rationales are provided later. Care planning should be individualized, based on the child's

and family's needs. Refer to Chapter 14 for the nursing process for pain management and to Chapter 11 for nursing interventions related to interrupted family processes. Additional information will be included later in the chapter as it relates to nursing management of children with specific disorders, as well as particular nursing interventions for deficient knowledge.

• • • ATRAUMATIC CARE • • •

When a child is diagnosed with congenital heart disease, involve the child life specialist early in the course of treatment. The child will likely have been undergoing diagnostic procedures such as electrocardiograms and echocardiograms, as well as open heart surgery. The child life specialist can be very helpful with providing atraumatic care.

Nursing Analysis

Decreased cardiac output related to structural defect, congenital anomaly, or ineffective heart pumping as evidenced by arrhythmias, edema, murmur, abnormal heart rate, or abnormal heart sounds

Goal/Outcome

Child or infant will demonstrate adequate cardiac output as evidenced by elastic skin turgor, brisk capillary refill, demonstrate pink color, pulse, and BP within normal limits for age, regular heart rhythm, adequate urinary output.

Increasing Cardiac Output (interventions with rationale)

- Monitor vital signs closely, especially BP and heart rate, *to detect increases or decreases.*
- Monitor cardiac rhythm via cardiac monitor *to detect arrhythmias quickly.*
- Observe for signs of hypoxia such as tachypnea, cyanosis, tachycardia, bradycardia, dizziness, and/or restlessness *to identify this change early.*
- Administer oxygen as needed *to correct hypoxia.*
- Place child in knee-to-chest or squatting position as needed *to increase systemic vascular resistance.*
- Administer antiarrhythmics, vasopressors, angiotensin-converting enzyme (ACE) inhibitors, beta-blockers, corticosteroids, or diuretics as prescribed *to improve cardiac output.*
- Monitor for signs of thrombosis such as restlessness, seizure, coma, oliguria, anuria, edema, hematuria, or paralysis *to identify this condition early.*
- Administer adequate hydration *to decrease possibility of thrombosis formation.*
- Cluster nursing care and other activities *to allow adequate periods of rest.*
- Anticipate child's needs *to decrease the child's stress, thereby decreasing oxygen consumption requirement.*

Nursing Analysis

Excess fluid volume related to compromised regulatory mechanism (ineffective cardiac muscle function) as evidenced by weight gain, edema, jugular vein distention, dyspnea, or adventitious breath sounds

Goal/Outcome

Child will attain appropriate fluid balance, will lose weight (fluid), edema or bloating will decrease, lung sounds will be clear, and heart sounds will be normal.

Encouraging Fluid Loss (interventions with rationale)

- Weigh daily on the same scale in a similar amount of clothing; *in children, weight is the best indicator of changes in fluid status.*
- Monitor location and extent of edema (measure abdominal girth daily if ascites is present); *a decrease in edema indicates positive increase in oncotic pressure.*
- Protect edematous areas from skin breakdown; *edema leads to increased risk for alterations in skin integrity.*
- Auscultate lungs carefully *to identify crackles, indicating pulmonary edema.*
- Assess work of breathing and respiratory rate; *increased work of breathing is associated with pulmonary edema.*
- Assess heart sounds for gallop; *the presence of S₃ may indicate fluid overload.*
- Maintain fluid restriction as ordered *to decrease intravascular volume and workload on the heart.*
- Strictly monitor intake and output *to quickly note discrepancies and provide intervention.*
- Provide sodium-restricted diet as ordered; *restricting sodium intake allows better kidney excretion of extra fluid.*
- Administer diuretics as ordered, and monitor for adverse effects *to encourage excretion of fluid and elimination of edema, reduce cardiac filling pressures, and increase kidney blood flow. Adverse effects include electrolyte imbalance as well as orthostatic hypotension.*

Nursing Analysis

Activity intolerance related to imbalanced oxygen supply and demand (ineffective cardiac muscle function, increased energy expenditure) as evidenced by exertional discomfort (squatting position), exertional dyspnea, weakness, or fatigue

Goal/Outcome

Child will increase activity level as tolerated: Child participates in play and activities (specify particular activities and level as individualized for each child).

Promoting Activity (interventions with rationale)

- Assess level of fatigue and activity tolerance *to determine baseline for comparison.*

- Note extent of dyspnea, oxygen requirement, or color change with exertion *to provide baseline for comparison.*
- Cluster care activities, allowing rest periods in between, *to conserve child's energy.*
- Work with the parent and child to determine a mutually satisfactory daily schedule *to allow adequate rest and energy conservation.*
- Instruct family and child in prescribed activity restrictions *to prevent fatigue while allowing some activity.*
- In the infant, avoid long periods of crying or prolonged nipple feeding; *these expend excessive calories.*
- Provide neutral thermal environment *to avoid increased oxygen and energy needs associated with excessive heat or cold.*

Nursing Analysis

Imbalanced nutrition (less than body requirements) related to the inability to increase adequate calories (due to increased energy expenditure and fatigue) as evidenced by food intake less than recommended daily allowance, weight loss or length/height and weight below accepted standards

Goal/Outcome

Child will improve nutritional intake, resulting in steady increase in weight and length/height and will feed without tiring easily.

Promoting Adequate Nutrition (interventions with rationale)

- Determine body weight and length/height norm for age *to determine a goal to work toward.*
- Assess child for food preferences that fall within dietary restrictions; *the child will be more likely to consume adequate amounts of foods that they like.*
- Weigh the child daily or weekly (according to health care provider order or institutional standard), and measure length/height weekly *to monitor for increased growth.*
- Offer highest-calorie meals at the time of day when the child's appetite is the greatest *to increase likelihood of increased caloric intake.*
- Provide increased-calorie shakes or puddings within diet restriction; *high-calorie foods increase weight gain.*
- Consult with the pediatric dietician *to provide optimal caloric intake within dietary restrictions.*
- Provide small, frequent feedings *to discourage tiring with feeding.*
- Feed infants with special nipple as needed *to decrease amount of energy expended for sucking.*
- Administer vitamin and mineral supplements as prescribed *to attain/maintain vitamin and mineral balance in the body.*

Nursing Analysis

Delayed development risk related to congenital disorder or chronic illness (effects of cardiac disease and necessary treatments, inadequate nutrition, or frequent separation from caregivers secondary to illness)

Goal/Outcome

Child will display development appropriate for age with evidence of cognitive and motor function within normal limits (individualized for each child)

Promoting Appropriate Development (interventions with rationale)

- Promote adequate caloric intake *to stimulate growth and provide adequate energy.*
- Provide age-appropriate developmental activities *to stimulate development.*
- Consult with the physical or occupational therapist or child life specialist *to determine activities most appropriate for the child within the constraints of the child's illness.*
- Schedule daily activities to allow for essential rest periods *for energy conservation.*
- Encourage parents, teachers, and playmates to be sensitive to child's self-image, using positive comments *to improve the child's self-concept.*
- As energy allows, encourage participation in all activities as feasible *to allow the child to feel normal.*

Based on your top three issues for Logan, describe appropriate nursing interventions.

Cardiac Catheterization

Nursing management of the child undergoing cardiac catheterization includes preprocedure nursing assessment and preparation of the child and family, postprocedural nursing care, and discharge teaching.

Assessment Before the Procedure

Obtain a thorough history and physical examination to establish a baseline. Measure vital signs. Note fever or other signs and symptoms of infection, which may necessitate rescheduling the procedure. Obtain the child's height and weight to aid in determining medication dosages. Assess the child for any allergies, especially to iodine and shellfish, because some contrast materials contain iodine as a base. Review the child's medications; medications such as anticoagulants are typically withheld for several days or longer prior to the procedure to reduce the child's risk for bleeding. Check the results of any laboratory tests, such as hemoglobin and hematocrit levels.

Perform a complete physical examination. Pay particular attention to assessing the child's peripheral pulses,

including pedal pulses. Use an indelible pen to mark the location of the child's pedal pulses so they can be easily assessed after the procedure. Document the location and quality in the child's medical record.

Educating the Child and Family Before the Procedure

Teach the parents and, if age-appropriate, the child, about all aspects of the procedure in order to decrease their anxiety. Let them know the procedure is commonly performed on an outpatient basis but that some health care providers or nurse practitioners require the child to be admitted for an overnight stay for observation. Include information about what the procedure involves, how long it will take, and any special instructions from the health care provider or nurse practitioner. Use a variety of teaching methods as appropriate, such as videos, books, and pamphlets.

Adapt these teaching methods to the child's developmental stage. For example, introduce the younger child to equipment through play therapy. For school-age and older children and their parents, offer a tour of the cardiac catheterization laboratory. Mention sounds and sights they may experience during the procedure. Explain the use of intravenous (IV) fluid therapy, sedation, and, if ordered, anesthesia to the child and parents. Tell the child that they may feel a sensation of the heart racing when the catheter is inserted. Also warn the older child that they may experience a feeling of warmth or stinging when the contrast material is injected. Encourage the child to use familiar ways to relax. If necessary, teach the child simple relaxation measures.

Tell families to withhold food and fluid for 4 to 6 hours before the procedure (as ordered). The parent should administer prescribed medications with a sip of water. On the day of the procedure, check to ensure that a signed informed consent form is on the child's medical record and that all necessary assessment data have been included. Just before the procedure, ask the child to void, and administer a sedative, as ordered. If appropriate and permitted, allow the parents to accompany the child to the catheterization area.

Teach the child and family what to expect after the procedure is completed. Inform the parents of the possible complications that might occur, such as bleeding, low-grade fever, loss of pulse in the extremity used for the catheterization, and development of **arrhythmia** (abnormal heart rhythm). Explain to the child that they will have a dressing over the catheter site and that they will need to keep the leg straight for several hours after the procedure. Teach the child and parent that frequent monitoring will be required after the procedure.

Assessment After the Procedure

Throughout the postprocedure period, closely monitor the child for complications of bleeding, arrhythmia, hematoma, and thrombus formation and infection. Evaluate

the child's vital signs, the neurovascular status of the lower extremities, and the pressure dressing over the catheterization site every 15 minutes for the first hour and then every 30 minutes for 1 hour. Vital signs should remain within acceptable parameters. Hypotension may signify hemorrhage due to perforation of the heart muscle or bleeding from the insertion site. Expect to monitor cardiac rhythm and oxygen saturation levels via pulse oximetry for the first few hours after the procedure to help identify possible complications.

Assess the child's distal pulses bilaterally for presence and quality. The pulse of the affected extremity may be slightly less than that of the other extremity in the initial postprocedure period, but it should gradually return to baseline. Also assess the color and temperature of the extremity; pallor or blanching would indicate an obstruction in blood flow. Check capillary refill and sensation to evaluate blood flow to the area.

Nursing Interventions Following Cardiac Catheterization

Maintain bed rest in the immediate postprocedure period. Ensure that the child maintains the extremity in a straight position for approximately 4 to 8 hours, depending on the approach used and the facility's policy. Inspect the pressure dressing frequently. Check to make sure that it is dry and intact, without evidence of bleeding. Reinforce the dressing as necessary and report any evidence of drainage on the dressing. If there is a risk of the dressing becoming soiled or wet, cover it with plastic.

TAKE NOTE!

If bleeding occurs after a cardiac catheterization, apply pressure 1 in above the site to create pressure over the vessel, thereby reducing the blood flow to the area.

Monitor the child's intake and output closely to ensure adequate hydration. The contrast material has a diuretic effect, so assess the child for signs and symptoms of dehydration and hypovolemia. Typically, the child resumes oral intake as tolerated, beginning with sips of clear liquids and progressing to their preprocedure diet. Continue IV fluids as ordered, and encourage oral fluid intake as allowed and ordered to promote elimination of the contrast material. Allow the child to talk about the experience and how and what they felt. Provide positive reinforcement for the child's actions.

Educating About Home Care Following Cardiac Catheterization

Provide child and family education before the child is discharged home (see Teaching Guidelines 19.1). Areas

TEACHING GUIDELINES 19.1 Providing Care After a Cardiac Catheterization

- Change the pressure dressing on the day after the procedure. Apply a dry sterile dressing or adhesive bandage for the next several days. Keep the dressing dry; cover it with plastic if there is a chance that the dressing could become wet or soiled.
- When changing the dressing, inspect the insertion site for redness, irritation, swelling, drainage, and bleeding. Report any of these to the health care provider or nurse practitioner.
- Check the temperature, color, sensation, and pulses on the child's extremities and compare. Report any changes to the health care provider or nurse practitioner.
- Resume the child's usual diet after the procedure; report any nausea or vomiting.
- Check the child's temperature at least once a day for approximately 3 days after the procedure. Report any temperature elevation of 100.4°F or greater.
- Avoid giving the child a tub bath for approximately 3 days after the procedure; use sponge baths or showers instead.
- Discourage strenuous exercise or activity for approximately 3 days after the procedure.
- Watch for changes in the child's appearance, such as changes in skin color, reports of the heart "fluttering" or "skipping a beat," fever, or difficulty breathing.
- Give acetaminophen (Tylenol) or ibuprofen (Motrin) for complaints of pain.
- Schedule a follow-up appointment with the health care provider or nurse practitioner in the time specified.

Based on KidsHealth Medical Experts. (2023). *Cardiac catheterization*. <https://kidshealth.org/en/parents/cardiac-catheter.html>; UCSF Benioff Children's Hospital. (2024). *Cardiac catheterization*. https://www.ucsfbenioffchildrens.org/education/cardiac_catheterization/

to address include site care, signs and symptoms of complications (especially within 24 hours after the catheterization, such as fever; bleeding or bruising at the catheterization site; or changes in color, temperature, or sensation in the extremity used), diet, and activity level.

THINKING ABOUT DEVELOPMENT

Jeremy Titus is a 2-year-old with congenital heart disease. He is having a cardiac catheterization today. How will the nurse encourage Jeremy to stay in bed and keep his leg straight for several hours following the catheterization? What types of activities would be appropriate for occupying Jeremy while he is confined to bed? How would the nurse's approach differ if Jeremy were an older child?

CONGENITAL HEART DEFECTS

In North America, more than 1% of newborn infants have CHD resulting from numerous causes. The prevalence of CHD ranges from six to 13 per 1,000 live births; premature infants have a higher rate (Altman, 2022). About one third of infants with CHD will have disease serious enough to result in death or will require cardiac catheterization or cardiac surgery within the first year of life. Complications of CHD include heart failure, hypoxemia, growth delay, developmental delay, and pulmonary vascular disease. Children with severe anomalies frequently experience failure to thrive.

With advances in palliative and corrective surgery over the past 60 years, many more children are now able to survive into adulthood. About 90% of children with CHD grow to be adults (Jone et al., 2022). As compared to healthy children, children with CHD tend to have poorer health overall and more frequently have additional morbidities either physical or neurodevelopmental in nature (Centers for Disease Control and Prevention [CDC], 2022). Due to the potential long-term effects that CHD may have on these children, nurses must be expertly equipped to care for them.

Pathophysiology

The exact cause of CHD is unknown. However, the belief is that it results from the interplay of several factors, including genetics (e.g., chromosomal alterations) and exposure during pregnancy to environmental factors (e.g., toxins, infections, chronic illnesses, and alcohol).

CHDs result from some interference in the development of the heart structure during fetal life. Subsequently, the septal walls or valves may fail to develop completely, or vessels or valves may be stenotic, narrowed, or transposed. Structures that formed to allow fetal circulation may fail to close after birth, altering the pressures necessary to maintain adequate blood flow.

After birth, with the change from fetal to newborn circulation, pressures within the chambers of the right side of the heart are less than those of the left side, and pulmonary vascular resistance is less than that for the systemic circulation. These normal pressure gradients are necessary for adequate circulation to the lungs and the rest of the body. However, these pressure gradients become disrupted if a structure has failed to develop, a fetal structure has failed to close, or a narrowing, stenosis, or transposition of a vessel has occurred. For example, blood typically flows from an area of higher pressure to one of lower pressure. If the ductus arteriosus fails to close, blood will move from the aorta to the pulmonary artery, ultimately increasing right atrial pressure. With this shunting of blood, highly oxygenated blood can mix with less oxygenated blood, interfering with the amount

available to the tissues via the systemic circulation. Some of the defects may result in significant hypoxemia, the sequelae of which include clubbing, **polycythemia** (excess amount of red blood cells [RBCs]), exercise intolerance, hypercyanotic spells, brain abscess, and cerebrovascular accident (CVA) (Jones et al., 2022; Schneider, 2023).

CHDs are categorized based on hemodynamic characteristics (blood flow patterns in the heart):

- Disorders with decreased pulmonary blood flow: tetralogy of Fallot and tricuspid atresia
- Disorders with increased pulmonary blood flow: PDA, atrial septal defect (ASD), and ventricular septal defect (VSD)
- Obstructive disorders: coarctation of the aorta, aortic stenosis, and pulmonic stenosis
- Mixed disorders: transposition of the great arteries (TGA), total anomalous pulmonary venous return (TAPVR), truncus arteriosus, and hypoplastic left heart syndrome (HLHS)

Therapeutic Management

Prenatal education about avoiding certain substances or preventing infection is essential to promote optimal outcomes for the fetus. Encourage parents of children with CHD to receive genetic counseling because of the probability of having subsequent children with a CHD. Children with small septal defects are urged to lead normal lives and often require no medical intervention. Therapeutic management of other forms of CHDs focuses on palliative care or a surgical corrective approach necessary for most of the

defects. In newborns and very young infants with severe cyanosis (tricuspid atresia, TGA), a prostaglandin infusion will maintain patency of the ductus arteriosus, improving pulmonary blood flow. Definitive correction of structural disorders requires surgical intervention. Table 19.2 describes the surgical procedures used for the various CHDs and the relevant nursing measures. Nursing management for the child with CHD will be provided following the disorders section.

Disorders With Decreased Pulmonary Blood Flow

Defects involving decreased pulmonary blood flow occur when there is some obstruction of blood flow to the lungs. As a result of the obstruction, pressure in the right side of the heart increases and becomes greater than that in the left side of the heart. Blood from the higher-pressure right side of the heart then shunts to the lower-pressure left side through a structural defect. Subsequently, deoxygenated blood mixes with oxygenated blood on the left side of the heart. This mixed blood, which is low in oxygen, is pumped via the systemic circulation to the body tissues.

Defects with decreased pulmonary blood flow are characterized by mild to severe oxygen desaturation. Typically, the child exhibits oxygen saturation levels ranging from 50% to 90%, which can produce severe cyanosis. To compensate for low blood oxygen levels, the kidneys produce the hormone erythropoietin to stimulate the bone marrow to produce more RBCs. This increase in RBCs is called polycythemia. Polycythemia can lead to an increase

TABLE 19.2 • Common Surgical Procedures and Nursing Measures for Congenital Heart Defects

Disorder	Surgical Procedure	Nursing Measures
Tetralogy of Fallot	Palliation with systemic-to-pulmonary anastomoses: <ul style="list-style-type: none"> • Blalock–Taussig shunt: an end-to-side anastomosis (or connection with a small Gore-Tex tube) of the subclavian artery and the pulmonary artery • Waterston shunt: anastomosis of the ascending aorta and the pulmonary artery • Definitive correction involves patch closure of the VSD and repair of the pulmonary valve and right ventricular outflow tract 	<ul style="list-style-type: none"> • Avoid BP measurements and venipunctures in the affected arm after a Blalock–Taussig shunt. Pulse will not be palpable in that arm because of use of the subclavian artery for the shunt. • Monitor for ventricular arrhythmias after corrective repair.
Tricuspid atresia	<ul style="list-style-type: none"> • Palliation with Blalock–Taussig shunt or pulmonary artery banding may be performed. • At 3–6 months of age, the superior vena cava is detached from the heart and connected to the pulmonary artery (Glenn procedure). • By age 2–5 years, a modified Fontan procedure may be performed. Systemic venous return is redirected to the pulmonary artery directly. 	<ul style="list-style-type: none"> • Monitor for atrial arrhythmias, left ventricular dysfunction, and protein-losing enteropathy. • Some children may eventually require a pacemaker.
Atrial septal defect (ASD)	<ul style="list-style-type: none"> • If small, the defect may be sutured closed. Larger defects may require a patch of pericardium or synthetic material. • Ostium secundum ASD may be repaired percutaneously via cardiac catheterization with a septal occluder. 	<ul style="list-style-type: none"> • Monitor for atrial arrhythmias (lifelong) after surgical closure. • With the septal occluders, strenuous activity should be avoided for 2 weeks after the procedure.^a

(continued)

TABLE 19.2 • Common Surgical Procedures and Nursing Measures for Congenital Heart Defects (continued)

Disorder	Surgical Procedure	Nursing Measures
Ventricular septal defect (VSD)	<ul style="list-style-type: none"> If surgical closure is required, it should be performed before permanent pulmonary vascular changes develop. Surgical closure may be in the form of suture closure of the VSD, transcatheter placement of a device in the defect, or Dacron patch closure. 	<ul style="list-style-type: none"> Monitor for ventricular dysrhythmias or atrioventricular block. With the clamshell occluding or Amplatzer device, strenuous activity should be avoided for 1 month after the procedure.^b
Atrioventricular canal defect	<ul style="list-style-type: none"> Pulmonary artery banding as palliation in very young infants Surgical correction by 3–18 months of age Patch closure of the septal defects and suturing of the valve leaflets or valve reconstruction are performed. 	<ul style="list-style-type: none"> Monitor for complete heart block postoperatively. Teach parents that mitral regurgitation is a long-term complication and may require valve replacement.
Patent ductus arteriosus (PDA)	<ul style="list-style-type: none"> PDA is closed by coil embolization or device via cardiac catheterization. May also be surgically ligated 	<ul style="list-style-type: none"> Monitor for bleeding and laryngeal nerve damage.
Coarctation of the aorta	<ul style="list-style-type: none"> Balloon angioplasty via cardiac catheterization is possible in some children. Most common surgical repair is resection of the narrowed portion of the aorta, followed by end-to-end reanastomosis. 	<ul style="list-style-type: none"> Preoperatively, administer prostaglandin medications as ordered to relax the ductal tissue. Postoperatively, measure and compare BP in all four extremities and quality of upper vs. lower pulses.
Aortic stenosis	<ul style="list-style-type: none"> Balloon dilation is accomplished via the umbilical artery in the newborn or the femoral artery via cardiac catheterization in the older child. 	<ul style="list-style-type: none"> Provide routine postcatheterization care. Teach parents that long-term aortic regurgitation requiring valve replacement may occur.
Pulmonic stenosis	<ul style="list-style-type: none"> Balloon dilation valvuloplasty is performed via cardiac catheterization to dilate the valve. This is effective in all but the most severe of cases, which will require surgical valvotomy. 	<ul style="list-style-type: none"> Provide routine postcatheterization care for balloon dilation. Explain to parents that prognosis is excellent.
Transposition of the great vessels (arteries)	<ul style="list-style-type: none"> Balloon atrial septotomy is usually done as soon as the diagnosis is made. A balloon-tipped catheter is passed through the atrial septum to enlarge the atrial septum. Surgical correction involves switching the arteries into their normal anatomic positions. 	<ul style="list-style-type: none"> Administer prostaglandin to maintain the open state of the ductus arteriosus, which will allow the mixing of poorly oxygenated blood with well-oxygenated blood. Monitor for rapid respirations and cyanosis. Administer oxygen as needed preoperatively.
Total anomalous pulmonary venous return	<ul style="list-style-type: none"> The pulmonary vein is repositioned to the back of the left atrium, and the ASD is closed. 	<ul style="list-style-type: none"> Monitor for dysrhythmias, heart block, and persistent heart failure.
Truncus arteriosus	<ul style="list-style-type: none"> VSD repair, separation of the pulmonary arteries from the aorta, with subsequent connection to the right ventricle with a valve conduit 	<ul style="list-style-type: none"> Preoperatively, administer prostaglandin infusion to prevent closing of the ductus arteriosus.
Hypoplastic left heart syndrome	<ul style="list-style-type: none"> Heart transplantation is the treatment of choice. Palliative staged treatment. First, Norwood procedure, reconstruction of the aorta and pulmonary arteries, includes a cardiac transplant. Second, bidirectional Glenn procedure, connection of the superior vena cava to the right pulmonary artery to increase the blood flow to the lungs. Third, modified Fontan procedure 	<ul style="list-style-type: none"> Preoperatively, administer prostaglandin infusion to prevent closing of the ductus arteriosus. After palliative repairs, monitor for dysrhythmias or worsening ventricular function.
Valve disorders	<ul style="list-style-type: none"> The incompetent valve is replaced with valve prosthesis. 	<ul style="list-style-type: none"> Lifelong anticoagulation therapy is necessary with prosthetic valves. Monitor prothrombin times. Monitor heart sounds for alterations.

^aCleveland Clinic. (2023). *Cardiac closure devices*. <https://my.clevelandclinic.org/health/treatments/16838-cardiac-implant-closure-devices-in-adults>^bAbbott. (2023). *Amplatzer septal occluder*. <https://www.myamplatzer.com/hcp/congenital-heart-defect-solutions/ventricular-septal-defects-vsd/>Based on Jone, P.-N., Kim, J. S., Burkett, D., Jacobsen, R., & VonAlvensleben, J., (2022). Cardiovascular disease. In M. Bunik, W. W. Hay, M. J. Levin, & M. J. Abzug (Eds.), *Current diagnosis and treatment: Pediatrics* (26th ed.). McGraw-Hill Education; Schneider, D. S. (2023). The cardiovascular system. In K. J. Marcante & R. M. Kliegman (Eds.), *Nelson's essentials of pediatrics* (8th ed.). Elsevier.

in blood volume and possibly blood viscosity, further taxing the workload of the heart. Although the number of RBCs increases, there is no change in the amount of blood that reaches the lungs for oxygenation. Disorders within this classification include tetralogy of Fallot and tricuspid atresia.

Tetralogy of Fallot

Tetralogy of Fallot is a CHD composed of four heart defects: pulmonic stenosis (a narrowing of the pulmonary valve and outflow tract, creating an obstruction of blood flow from the right ventricle to the pulmonary artery); VSD; overriding aorta (enlargement of the aortic valve to the extent that it appears to arise from the right and left ventricles rather than the anatomically correct left ventricle); and right ventricular hypertrophy (the muscle walls of the right ventricle increase in size due to continued overuse as the right ventricle attempts to overcome a high-pressure gradient). Surgical intervention is usually required during the first year of life (Jone et al., 2022; Schneider, 2023).

Pathophysiology

With pulmonic stenosis, the blood flow from the right ventricle is obstructed and slowed, resulting in a decrease in blood flow to the lungs for oxygenation and a decrease in the amount of oxygenated blood returning to the left atrium from the lungs. The obstructed flow also increases the pressure in the right ventricle. This blood, which is poorly oxygenated, is then shunted across the VSD into the left atrium. Poorly oxygenated blood also travels through the overriding aorta (if it extends to both ventricles). In some cases when the VSD is large, the pressure in the right ventricle may be equal to that of the left ventricle. In this case, the path of blood shunting depends on which circulation is exerting the higher pressure, pulmonary or systemic.

Regardless of which way shunting occurs, a mixing of oxygenated and poorly oxygenated blood occurs, with this blood ultimately being pumped into the systemic circulation. The oxygen saturation of the blood in the systemic circulation is reduced, leading to cyanosis. The degree of cyanosis depends on the extent of the pulmonic stenosis, the size of the VSD, and the vascular resistance of the pulmonary and systemic circulations.

Tetralogy of Fallot is usually diagnosed during the first few weeks of life due to the presence of a murmur and/or cyanosis. Some newborns may be acutely cyanotic, while others may exhibit only mild cyanosis that gradually becomes more severe, particularly during times of stress as the child grows older. Most often, infants with tetralogy of Fallot have a PDA at birth, providing additional pulmonary blood flow and thereby decreasing the severity of the initial cyanosis. Later, as the ductus arteriosus closes, such as within the first few days of life, more severe cyanosis can occur (Fig. 19.2) (Jone et al., 2022; Schneider, 2023).

Nursing Assessment

Nursing assessment consists of the health history, physical examination, and laboratory and diagnostic tests.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Obtain the health history, noting a history of color changes associated with feeding, activity, or crying. Determine if the infant or child is demonstrating hypercyanotic spells. Hypercyanosis develops suddenly and is manifested as increased cyanosis, hypoxemia, dyspnea, and agitation. If the infant's oxygen demand is greater than the supply, such as with crying or during feeding, then the spell progresses to anoxia. When the degree of cyanosis is severe and persistent, the infant may become unresponsive. As the infant gets older, they may use specific postures, such as bending at the knees or assuming the fetal position, to relieve a hypercyanotic spell. The walking infant or toddler may squat periodically. These positions improve pulmonary blood flow by increasing systemic vascular resistance. Ask the parents if they have noticed any of these unusual positions. Note history of irritability, sleepiness, or difficulty breathing.

During the physical examination, observe the skin color and note any evidence of cyanosis. Also observe for changes in skin color with positional changes, and inspect the fingers for clubbing. Note if the child has a hypercyanotic spell during the assessment. Count the child's respiratory rate and observe work of breathing, noting retractions, shortness of breath, or noisy breathing. Document oxygen saturation via pulse oximetry; it will likely be decreased. Auscultate the chest for adventitious breath sounds, which may suggest the development of heart failure. Auscultate the heart, noting a loud, harsh murmur characteristic of this disorder.

LABORATORY AND DIAGNOSTIC TESTS

Note increased hematocrit, hemoglobin, and RBC count associated with polycythemia. Additional testing may include:

- **Echocardiography** (ultrasound study of structure and motion of heart), possibly revealing right ventricular hypertrophy, decreased pulmonary blood flow, and reduced size of the pulmonary artery
- Electrocardiogram (ECG), indicating right ventricular hypertrophy
- Cardiac catheterization and angiography, which reveal the extent of the structural defects

Tricuspid Atresia

Tricuspid atresia is a CHD in which the valve between the right atrium and right ventricle fails to develop. As a result, there is no opening to allow blood to flow from the right atrium to the right ventricle and subsequently through the pulmonary artery into the lungs (Jone et al., 2022; Schneider, 2023).

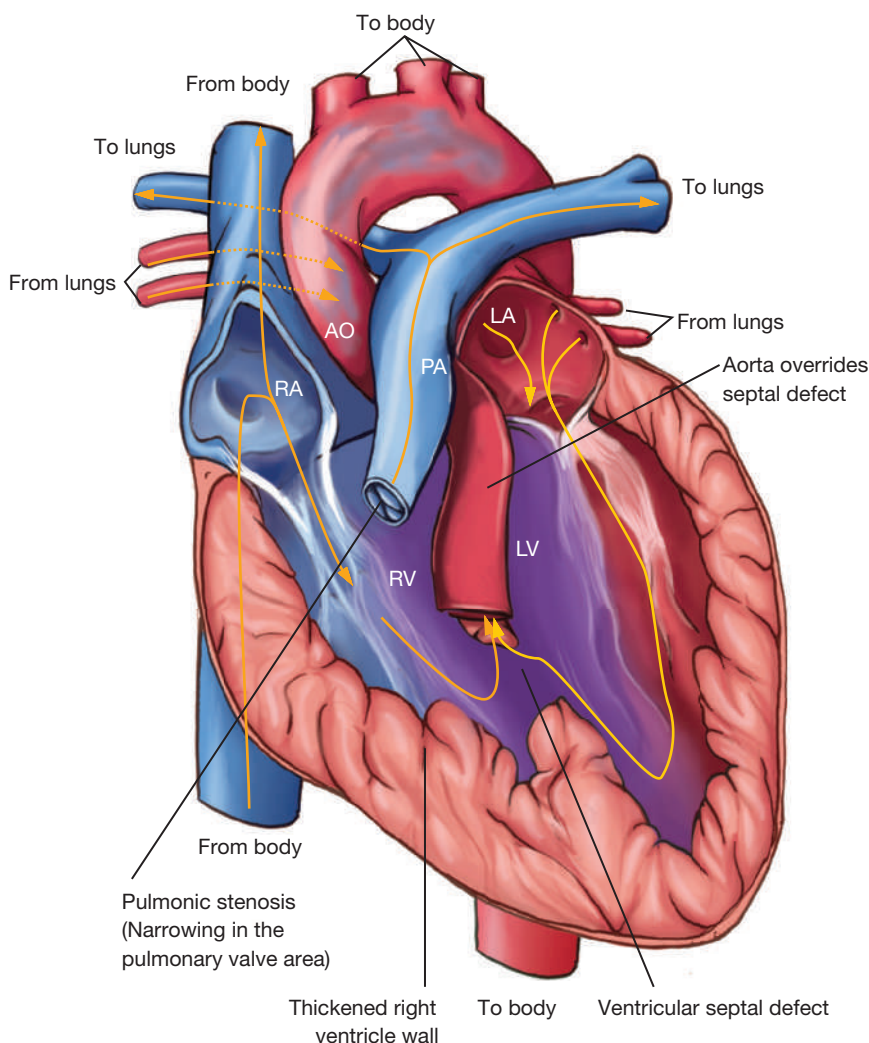


FIGURE 19.2 Tetralogy of Fallot.

CONSIDER THIS!

Ava Gardener, 2 weeks old, is brought to the clinic by her parent. She presents with trouble feeding. Her parent states, “When Ava eats, she seems to have trouble breathing, and a couple of times she has looked a little bluish.” As the nurse takes Ava into her arms, Ava has a hypercyanotic spell.

Ava is to be admitted to the hospital secondary to suspected tetralogy of Fallot. Ava’s parent is very upset about the diagnosis. They say, “My poor baby, she’ll never ever be able to run and play like a normal child.”

How should the nurse respond? How would you feel if your young baby was diagnosed with a serious disorder? What type of support can the nurse provide to Ava’s parent?

Pathophysiology

In tricuspid atresia, blood returning from the systemic circulation to the right atrium cannot directly enter the right ventricle due to agenesis of the tricuspid valve. Subsequently, deoxygenated blood may pass through an opening in the atrial septum (patent foramen ovale) into

the left atrium, never entering the pulmonary vasculature. Thus, deoxygenated blood mixes with oxygenated blood in the left atrium. The blood then travels to the lungs through a PDA. Most cases of tricuspid atresia are associated with a VSD, and the newborn receives inadequately oxygenated blood (Fig. 19.3) (Jone et al., 2022; Schneider, 2023).

Nursing Assessment

Nursing assessment consists of the health history, physical examination, and laboratory and diagnostic tests.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Note the infant’s history since birth. Document history of cyanosis either at birth or a few days later when the ductus arteriosus closed. Note history of rapid respirations and difficulty with feeding. Inspect the skin for cyanosis or a pale gray color. Observe the apical impulse, noting overactivity. Evaluate the baby’s sucking strength (will usually have a weak or poor suck). Count the respiratory rate, noting tachypnea. Note increased work

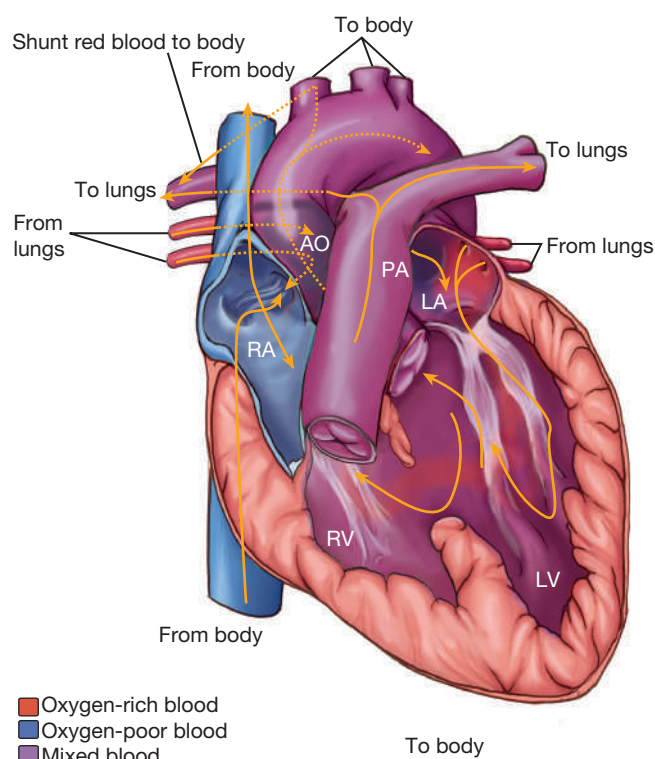


FIGURE 19.3 Tricuspid atresia.

of breathing. Auscultate the lungs, noting crackles or wheezes if heart failure is beginning to develop. Auscultate the heart, noting a murmur. Palpate the skin, noting coolness and clamminess of the extremities. Document the presence of clubbing in the older infant or child.

LABORATORY AND DIAGNOSTIC TESTING

Laboratory and diagnostic testing is similar to that for tetralogy of Fallot. A complete blood cell (CBC) count is needed to assess compensatory increases in hematocrit, hemoglobin, and erythrocyte (RBC) count, indicating the development of polycythemia. Pulse oximetry or arterial blood gas tests may be used to determine oxygen saturation levels (typically reduced). Additional testing may include:

- Echocardiography, revealing absence of tricuspid valve or underdeveloped right ventricle
- ECG, indicating possible heart failure
- Cardiac catheterization and angiography, which reveal the extent of the structural defects

Disorders With Increased Pulmonary Flow

Most CHDs involve increased pulmonary blood flow. Normally, the left side of the heart has a higher pressure than the right side. Defects with connections involving the left and right sides will shunt blood from the

higher-pressure left side to the lower-pressure right side. Even a small pressure gradient such as a 1- to 3-mm difference between the left and the right sides will produce a movement of blood from the left to the right. In turn, the increase in blood on the right side of the heart will cause a greater amount of blood to move through the heart. If the amount of blood flowing to the lungs is large, the child may develop heart failure early in life. In addition, right ventricular hypertrophy may result. Sometimes, with ventricular hypertrophy, the right side of the heart pumps so forcefully that left-to-right shunting is reversed to right-to-left shunting. If this occurs, deoxygenated blood mixes with oxygenated blood, thereby lowering the overall blood oxygen saturation level.

Excessive blood flow to the lungs can produce a compensatory response such as tachypnea or tachycardia. Tachypnea increases caloric expenditure; poor cellular nutrition from decreased peripheral blood flow leads to feeding problems. Subsequently, the infant experiences poor weight gain, which delays overall growth and development. Increased pulmonary blood flow results in decreased systemic blood flow, so sodium and fluid retention may occur. Increased pulmonary blood flow also places the child at higher risk for pulmonary infections. As the child grows, the continuous increased pulmonary blood flow will cause vasoconstriction of the pulmonary vessels, actually decreasing the pulmonary blood flow. This may lead to pulmonary hypertension. For children with congenital defects with increased pulmonary blood flow, oxygen supplementation is not helpful. Oxygen acts as a pulmonary vasodilator. If pulmonary dilation occurs, pulmonary blood flow is even greater, causing tachypnea, increasing lung fluid retention, and eventually causing a much greater problem with oxygenation. Over time, continuous increased pulmonary blood flow may cause pulmonary vasoconstriction and pulmonary hypertension. Therefore, preventing the development of pulmonary disease via early surgical correction is essential.

Examples of defects with increased pulmonary blood flow are ASD, VSD, atrioventricular canal defect, and PDA.

Atrial Septal Defect

An ASD is a passageway or hole in the wall (septum) that divides the right atrium from the left atrium (Fig. 19.4). Three types of ASDs are identified based on the location of the opening:

- Ostium primum (ASD1): The opening is at the lower portion of the septum.
- Ostium secundum (ASD2): The opening is near the center of the septum.
- Sinus venosus defect: The opening is near the junction of the superior vena cava and the right atrium.

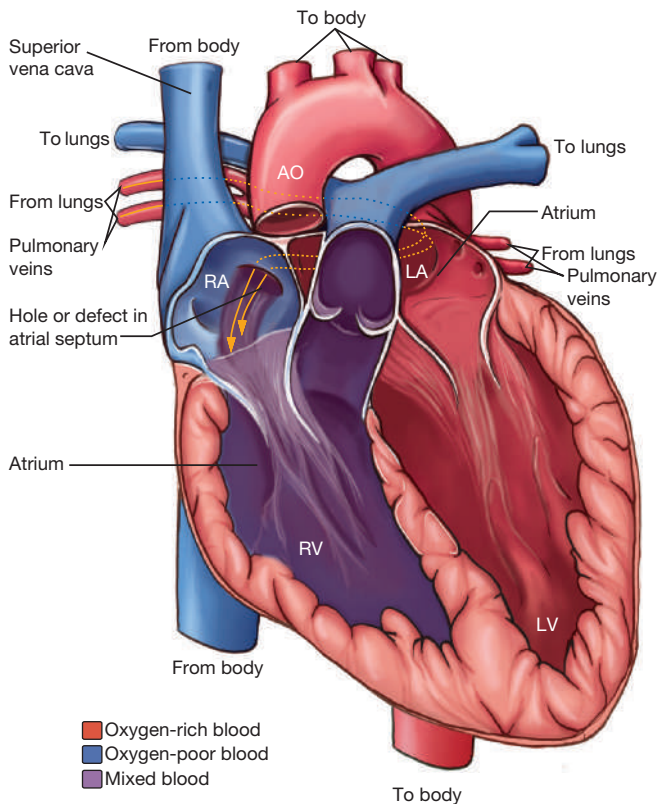


FIGURE 19.4 Atrial septal defect; note the opening between the two atria.

When the ASD is small, most infants may have a spontaneous closure within the first 18 months of life. If it does not spontaneously close by age 3, the child will most likely need corrective surgery (Jone et al., 2022; Schneider, 2023).

Pathophysiology

With ASD, blood flows through the opening from the left atrium to the right atrium due to pressure differences. The shunting increases the blood volume entering the right atrium. This, in turn, leads to increased blood flow into the lungs. If untreated, the defect can cause problems such as pulmonary hypertension, heart failure, atrial arrhythmias, or stroke (Jone et al., 2022; Schneider, 2023).

Nursing Assessment

Most children with ASDs are asymptomatic. However, a very large defect can cause increased blood flow, leading to heart failure, which results in shortness of breath, easy fatigability, or poor growth.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Obtain the health history, noting poor feeding as an infant, decreased ability to keep up with peers, or history of difficulty growing. Observe the child's chest, noting a

hyperdynamic precordium. Auscultate the heart, noting a fixed split-second heart sound and a systolic ejection murmur, best heard in the pulmonic valve area. Palpate along the left sternal border for a right ventricular heave.

LABORATORY AND DIAGNOSTIC TESTS

Echocardiography is done to confirm the diagnosis. An **electrocardiogram** (graphic recording of the heart's electrical activity) may show normal sinus rhythm or prolonged PR intervals. The chest radiograph may show enlargement of the heart and increased vascularity of the lungs.

Ventricular Septal Defect

A VSD is an opening between the right and left ventricular chambers of the heart (Fig. 19.5). It is one of the most common CHDs and accounts for about 30% of all CHDs. Spontaneous closure of small VSDs occurs in about half of children by age 2 years. Long-term outcomes for surgically repaired VSDs are good. Repair of larger defects by 2 years of age is recommended to prevent the development of pulmonary vascular disease (Jone et al., 2022; Schneider, 2023).

Pathophysiology

In VSD, there is an abnormal opening between the right and the left ventricles. The opening varies in size, from

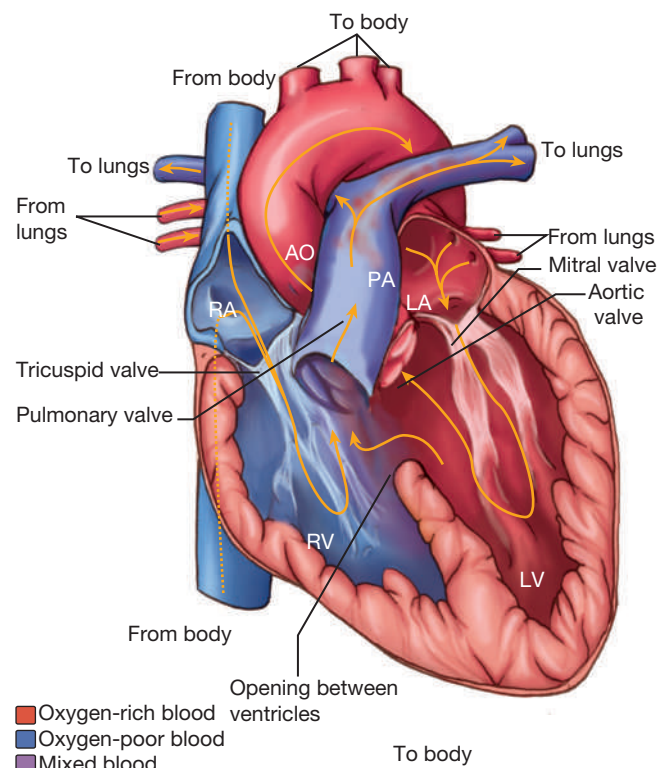


FIGURE 19.5 Ventricular septal defect; note the opening between the ventricles.

as small as a pinhole to a complete opening between the ventricles so that the right and the left sides are as one. Children with small VSDs may remain asymptomatic. In other children, blood shunts across the opening in the septum. Pulmonary vascular resistance and systemic vascular resistance determine the direction of blood flow. A left-to-right shunt results when pulmonary vascular resistance is low. Increased amounts of blood flowing into the right ventricle are then pumped to the pulmonary circulation, eventually causing an increase in pulmonary vascular resistance. Increased pulmonary vascular resistance leads to increased pulmonary artery pressure (pulmonary hypertension) and right ventricular hypertrophy. When the pulmonary vascular resistance exceeds the systemic vascular resistance, right-to-left shunting of blood across the VSD occurs, resulting in Eisenmenger syndrome (pulmonary hypertension and cyanosis). Heart failure commonly occurs in children with moderate to severe unrepaired VSDs. Children with VSDs are also at risk for the development of aortic valve regurgitation as well as infective endocarditis (Jone et al., 2022; Schneider, 2023).

Nursing Assessment

Initially, the newborn may not exhibit any signs and symptoms at birth because left-to-right shunting is most likely minimal due to the high pulmonary resistance common after birth.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Determine the health history, which commonly reveals signs of heart failure around 4 to 8 weeks of age. Note history of tiring easily, particularly with exertion or feeding. Document the child's growth history, noting difficulty thriving. Ask the parent about color change or diaphoresis with nipple feeding in the infant. Note history of frequent pulmonary infections, shortness of breath, and possibly edema. Inspect the extremities for edema, noting whether pitting is present. Note mild tachypnea.

Auscultate the heart, noting a characteristic holosystolic harsh murmur along the left sternal border. In some instances, a murmur may be noted only with excessive blood flow across the opening. Adventitious lung sounds may be auscultated if the child is experiencing heart failure. Palpate the chest for a thrill.

LABORATORY AND DIAGNOSTIC TESTS

Magnetic resonance imaging (MRI) or echocardiogram with color flow Doppler may reveal the opening as well as the extent of left-to-right shunting. These studies may also identify right ventricular hypertrophy and dilation of the pulmonary artery resulting from the increased blood flow. Cardiac catheterization may be used to evaluate the extent of blood flow being pumped to the pulmonary circulation and to evaluate hemodynamic pressures.

Atrioventricular Septal Defect

Atrioventricular septal defect (AVSD) accounts for 4% of CHD. Thirty-five to forty percent of children with Down syndrome and CHD have this defect (Jone et al., 2022).

Pathophysiology

AVSD occurs because of failure of the endocardial cushions to fuse (Fig. 19.6). These cushions are needed to separate the central parts of the heart near the tricuspid and mitral (AV) valves. The complete AVSD involves ASDs and VSDs as well as a common AV orifice and a common AV valve. Partial and transitional forms of AVSD also occur, involving variations of the complete form.

The complete AVSD permits oxygenated blood from the lungs to enter the left atrium and ventricle, crossing over the atrial or ventricular septum and returning to the lungs via the pulmonary artery. This recirculation problem, which typically involves a left-to-right shunt, is inefficient because the left ventricle must pump blood back to the lungs and also meet the body's peripheral demand for oxygenated blood. Subsequently, the left ventricle must pump two to three times more blood than in a normal heart. Therefore, this specific type of cardiac defect causes a large left-to-right shunt; an increased workload of the left ventricle; and high pulmonary arterial pressure, resulting in an increased amount of blood in the lungs and causing pulmonary edema (Jone et al., 2022; Schneider, 2023).

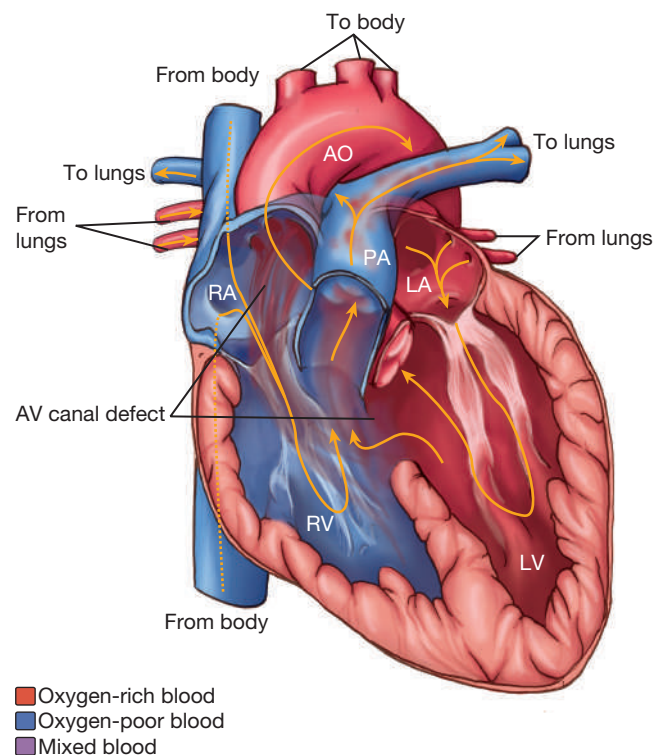


FIGURE 19.6 Atrioventricular canal defect.

Nursing Assessment

The infant with a complete AVSD commonly exhibits moderate to severe signs and symptoms of heart failure. However, for infants with a partial or transitional AVSD, the signs and symptoms will be subtler.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Obtain the health history, noting frequent respiratory infections and difficulty gaining weight. Ask the parent if the infant has been experiencing difficulty feeding or increased work of breathing.

Inspect the skin, fingernails, and lips for cyanosis. Observe for retractions, tachypnea, and nasal flaring. Auscultate the lungs and heart, noting rales and a loud murmur. The murmur is commonly noted within the first 2 weeks of life. Infants with a partial or transitional AVSD defect may display more subtle signs.

LABORATORY AND DIAGNOSTIC TESTS

Echocardiography will reveal the extent of the defect and shunting as well as right ventricular hypertrophy. ECG may indicate right ventricular hypertrophy and possible first-degree heart block due to impulse blocking before reaching the AV node.

Patent Ductus Arteriosus

PDA is failure of the ductus arteriosus, a fetal circulatory structure, to close within the first few weeks of life (Fig. 19.7).

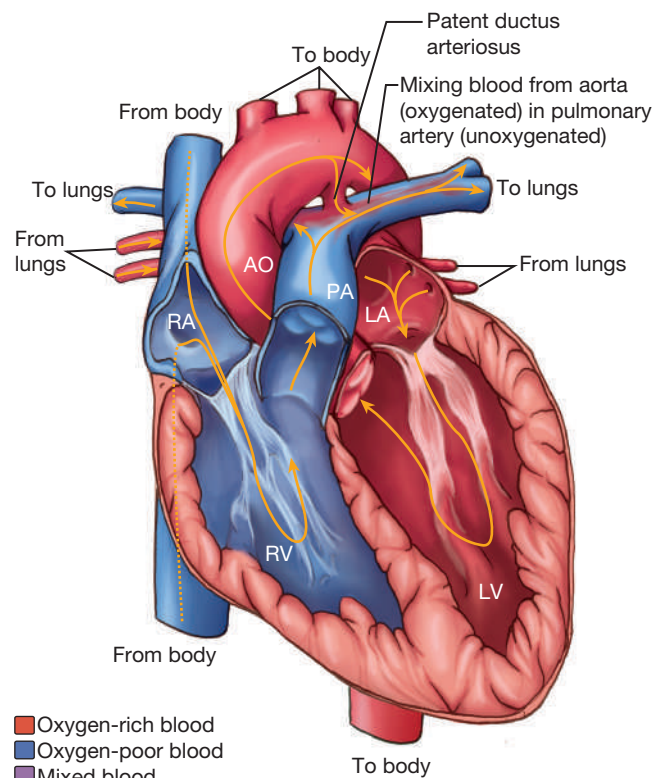


FIGURE 19.7 Patent ductus arteriosus.

As a result, there is a connection between the aorta and pulmonary artery. PDA is the second most common CHD and accounts for 10% of CHD cases (Jone et al., 2022). PDA occurs much more frequently in premature than in term infants and in infants born at high altitudes compared with those born at sea level. Infants with other CHDs that result in right-to-left shunting of blood and cyanosis may additionally display a PDA. In these infants, the PDA allows for some level of oxygenated blood to reach the systemic circulation (Jone et al., 2022; Schneider, 2023).

Pathophysiology

Failure of the ductus arteriosus to close leads to continued blood flow from the aorta to the pulmonary artery. Blood returning to the left atrium passes to the left ventricle, enters the aorta, and then travels to the pulmonary artery via the PDA instead of entering the systemic circulation. This altered blood flow pattern increases the workload of the left side of the heart. Pulmonary vascular congestion occurs, causing an increase in pressure. Right ventricular pressure increases in an attempt to overcome this increase in pulmonary pressure. Eventually, right ventricular hypertrophy occurs (Jone et al., 2022; Schneider, 2023).

Nursing Assessment

The symptoms of PDA depend on the size of the ductus arteriosus and the amount of blood flow it carries. If it is small, the infant may be asymptomatic. Some infants demonstrate signs and symptoms of heart failure.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Determine the health history, which may reveal frequent respiratory infections, fatigue, and poor growth and development. On physical examination, note tachycardia, tachypnea, bounding peripheral pulses, and a widened pulse pressure. The diastolic BP typically is low due to the shunting. Auscultate the lungs and heart, noting rales if heart failure is present. Note a harsh, continuous, machine-like murmur, usually loudest under the left clavicle at the first and second intercostal spaces.

LABORATORY AND DIAGNOSTIC TESTS

Echocardiogram reveals the extent of the defective opening and confirms the diagnosis. ECG may be normal, or it may indicate ventricular hypertrophy, especially if the defect is large. Chest radiography demonstrates cardiomegaly.

Obstructive Disorders

Another group of CHDs is classified as obstructive disorders. These disorders involve some type of narrowing of a major vessel, interfering with the ability of the blood

to flow freely through the vessel. As a result, peripheral circulation or blood flow to the lungs is affected. Increased pressure backing up toward the heart causes an increased workload on the heart. Examples of defects in this group include coarctation of the aorta, aortic stenosis, and pulmonic stenosis (PS).

Coarctation of the Aorta

Coarctation of the aorta is narrowing of the aorta, the major blood vessel carrying highly oxygenated blood from the left ventricle of the heart to the rest of the body (Fig. 19.8). It accounts for about 10% of CHDs (Schneider, 2023).

Pathophysiology

Coarctation of the aorta occurs most often in the area near the ductus arteriosus. The narrowing can be preductal (between the subclavian artery and the ductus arteriosus) or postductal (after the ductus arteriosus). As a result of the narrowing, blood flow is impeded, causing pressure to increase in the area proximal to the defect and to decrease in the area distal to it. Thus, BP is increased in the heart and the upper portions of the body and decreased in the lower portions of the body. Left ventricular afterload is increased, and in some children, this may lead to heart failure. Collateral circulation may also develop as the body attempts to ensure adequate

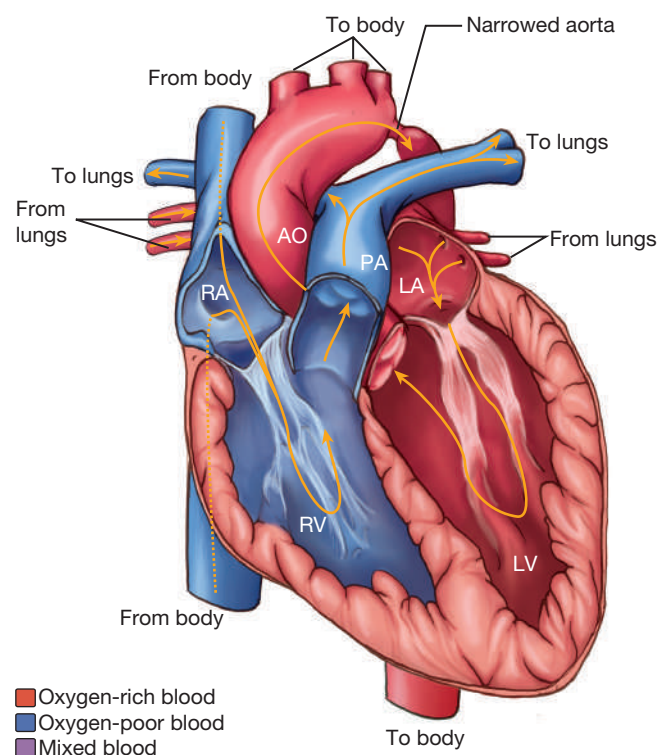


FIGURE 19.8 Coarctation of the aorta.

blood flow to the descending aorta. Due to the elevation in BP, the child is also at risk for aortic rupture, aortic aneurysm, and CVA (Jone et al., 2022; Schneider, 2023).

Nursing Assessment

The extent of the symptoms depends on the severity of the coarctation. Some children with coarctation of the aorta grow well into the school-age years before the defect is discovered.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Determine the health history, noting problems with irritability and frequent epistaxis. In older children, there may also be reports of leg pain with activity, dizziness, fainting, and headaches. Assess pulses throughout, noting full, bounding pulses in the upper extremities with weak or absent pulses in the lower extremities. Determine BP in all four extremities. BP in the upper extremities may be 20 mm Hg or higher than that in the lower extremities. Inspect the school-age child's chest, noting notching of the ribs. Auscultate the heart for a soft or moderately loud systolic murmur, most often heard at the base of the heart (on the back or in the left axilla) (Jone et al., 2022).

LABORATORY AND DIAGNOSTIC TESTS

Diagnosis of coarctation of the aorta is based primarily on the history and physical examination. In addition, an echocardiogram may disclose the extent of narrowing and evidence of collateral circulation. Chest radiography may reveal left-sided cardiac enlargement and rib notching, indicative of collateral arterial enlargement. Other tests, such as ECG, computed tomography, or MRI, may be done to provide additional evidence about the extent of the coarctation and subsequent effects.

Aortic Stenosis

Aortic stenosis is a condition causing obstruction of the blood flow between the left ventricle and the aorta. The incidence of aortic stenosis is about 5% of all CHDs (Schneider, 2023).

Pathophysiology

Aortic stenosis can be caused by a muscle obstruction below the aortic valve, an obstruction at the valve itself, or an aortic narrowing just above the valve (Fig. 19.9). The most common type is an obstruction of the valve itself, called aortic valve stenosis. The aortic valve consists of three very pliable leaflets. Normally, the leaflets of the aortic valve spread open easily when the left ventricle ejects blood into the aorta. Aortic stenosis occurs when the aortic valve narrows, causing an obstruction between the left ventricle and the aorta. As a result, cardiac output

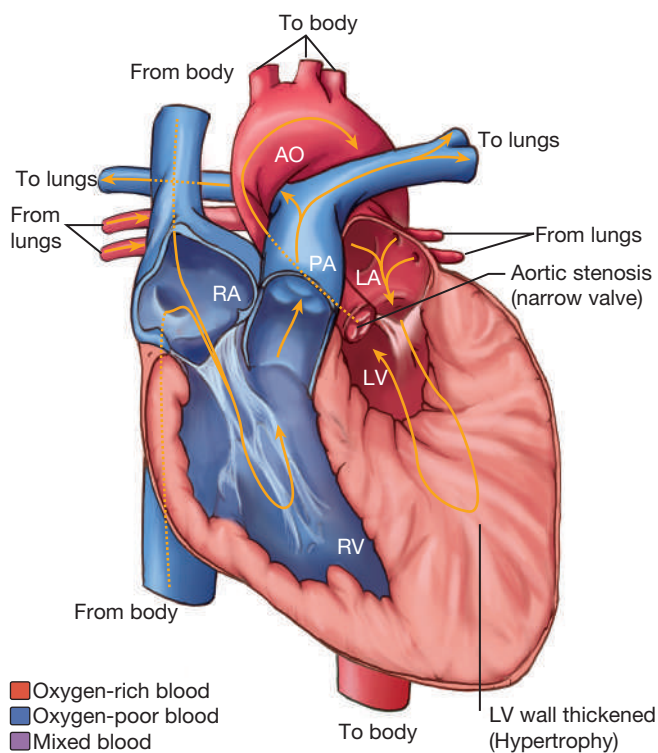


FIGURE 19.9 Aortic stenosis.

decreases. When the aortic valve does not function properly, the left ventricle must work harder to pump blood into the aorta. Because of the increased workload, the left ventricular muscle hypertrophies. If this continues, left ventricular failure can occur, leading to a backup of pressure in the pulmonary circulation and pulmonary edema. Heart failure may occur, but this is more commonly seen in the infant (Jones et al., 2022; Schneider, 2023).

Nursing Assessment

Typically, the child with aortic stenosis is asymptomatic. However, it is important to obtain an accurate health history and perform a physical examination.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Obtain the child's health history, noting easy fatigability or complaints of chest pain similar to anginal pain when active. Dizziness with prolonged standing may also be reported. In the infant, note difficulty with feeding. Palpate the child's pulse; if aortic stenosis is severe, the pulses may be faint. Palpate the child's chest, noting a thrill at the base of the heart. Auscultate the heart, noting a systolic murmur best heard along the left sternal border with radiation to the right upper sternal border.

LABORATORY AND DIAGNOSTIC TESTS

The echocardiogram is the most important noninvasive test to identify aortic stenosis. An ECG may be normal in

children with mild to moderate forms of aortic stenosis. For children with severe aortic stenosis, left ventricular hypertrophy may be determined from the ECG. For children experiencing easy fatigability and chest pain, an exercise stress test may be done to evaluate the degree of cardiac compromise.

Pulmonic Stenosis

PS is a condition that causes an obstruction in blood flow between the right ventricle and the pulmonary arteries. Pulmonic stenosis occurs in 0.6 to 0.8 per 1,000 live births (Peng, 2022). It is often associated with other heart anomalies and with genetic syndromes. Children may be asymptomatic, although some children with severe pulmonic stenosis may demonstrate cyanosis (Peng, 2022).

Pathophysiology

Pulmonic stenosis may occur as a muscular obstruction below the pulmonary valve, an obstruction at the valve, or a narrowing of the pulmonary artery above the valve (Fig. 19.10). Valve obstruction is the most common form of PS. Normally, the pulmonary valve is constructed with three thin and pliable valve leaflets; they spread apart easily, allowing the right ventricle to eject blood freely into the pulmonary artery. The most common problem

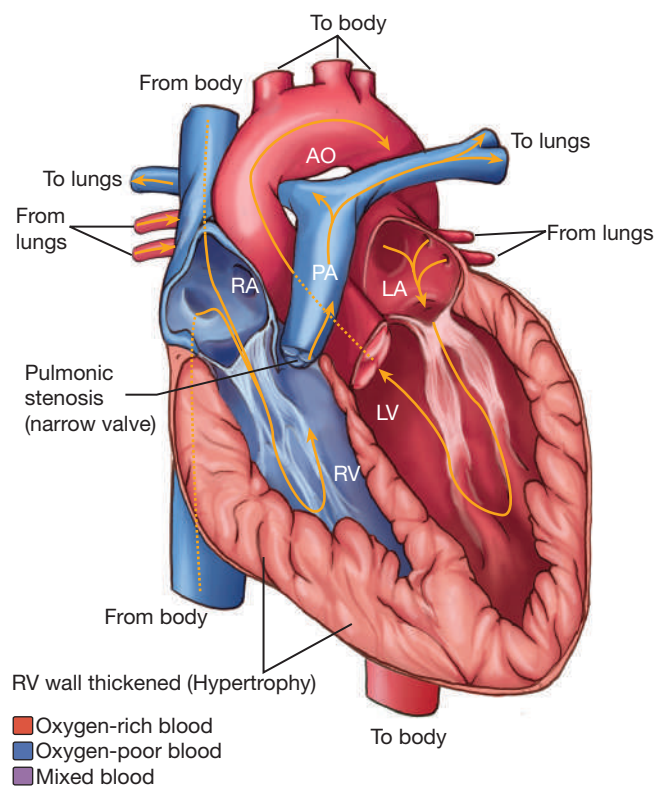


FIGURE 19.10 Pulmonic stenosis.

causing pulmonic stenosis is that the pulmonary valve leaflets are thickened and fused together along their separation lines, causing the obstruction to blood flow. The right ventricle has an additional workload, causing the muscle to thicken, resulting in right ventricular hypertrophy and decreased pulmonary blood flow. When the pulmonary valve is severely obstructed, the right ventricle cannot eject sufficient blood into the pulmonary artery. As a result, pressure in the right atrium increases, which could lead to a reopening of the foramen ovale. If this occurs, deoxygenated blood would pass through the foramen ovale into the left side of the heart and would then be pumped to the systemic circulation. In some cases, a PDA may be present, thus allowing for some compensation by shunting blood from the aorta to the pulmonary circulation for oxygenation (Peng, 2022).

Nursing Assessment

The child with pulmonic stenosis may be asymptomatic or may exhibit signs and symptoms of mild heart failure. If the stenosis is severe, the child may demonstrate cyanosis. Therefore, it is important for the nurse to obtain an accurate health history and physical examination.

HEALTH HISTORY AND PHYSICAL EXAMINATION

Elicit the health history, noting mild dyspnea or cyanosis with exertion. Document the child's growth history, which is typically normal. Carefully palpate the sternal border for a thrill (not always present). Auscultate the heart, noting a high-pitched click following the second heart sound and a systolic ejection murmur loudest at the upper left sternal border.

LABORATORY AND DIAGNOSTIC TESTS

An echocardiogram reveals the extent of obstruction present at the valve, as well as right ventricular hypertrophy. An ECG also helps to detect right ventricular hypertrophy.

Mixed Defects

Mixed defects are CHDs that involve a mixing of well-oxygenated blood with poorly oxygenated blood. As a result, systemic blood flow contains a lower oxygen content. Cardiac output is decreased, and heart failure occurs. Examples of mixed defects include TGA, total anomalous pulmonary venous connection (TAPVC), truncus arteriosus, and HLHS.

Transposition of the Great Arteries

TGA is a CHD in which the pulmonary artery and the aorta are transposed from their normal positions. The aorta arises from the right ventricle instead of the left ventricle, and the pulmonary artery arises from the left ventricle

instead of the right ventricle. TGA accounts about 5% of all CHD cases (Schneider, 2023). It is most often diagnosed in the first few days of life when the infant manifests cyanosis, which indicates decreased oxygenation. As the ductus arteriosus closes, the symptoms will worsen. Corrective surgery is usually performed by age 4 to 7 days.

PATHOPHYSIOLOGY

TGV creates a situation in which poorly oxygenated blood returning to the right atrium and ventricle is then pumped out to the aorta and back to the body (Fig. 19.11). Oxygenated blood returning from the lungs to the left atrium and ventricle is then sent back to the lungs through the pulmonary artery. Unless there is a connection somewhere in the circulation where the oxygen-rich and oxygen-poor blood can mix, all the organs of the body will be poorly oxygenated. Often, the ductus arteriosus remains patent, allowing for some mixing of blood. Similarly, if a VSD is also present, mixing of blood may occur, and cyanosis will be delayed. However, these associated defects can lead to increased pulmonary blood flow that increases pressure in the pulmonary circulation. This predisposes the child to heart failure (Jone et al., 2022; Schneider, 2023).

NURSING ASSESSMENT

Significant cyanosis without a murmur in the newborn period is highly indicative of TGA. In some infants, cyanosis will not develop until several days of age as the PDA closes. In infants with septal defects, cyanosis may be further delayed.

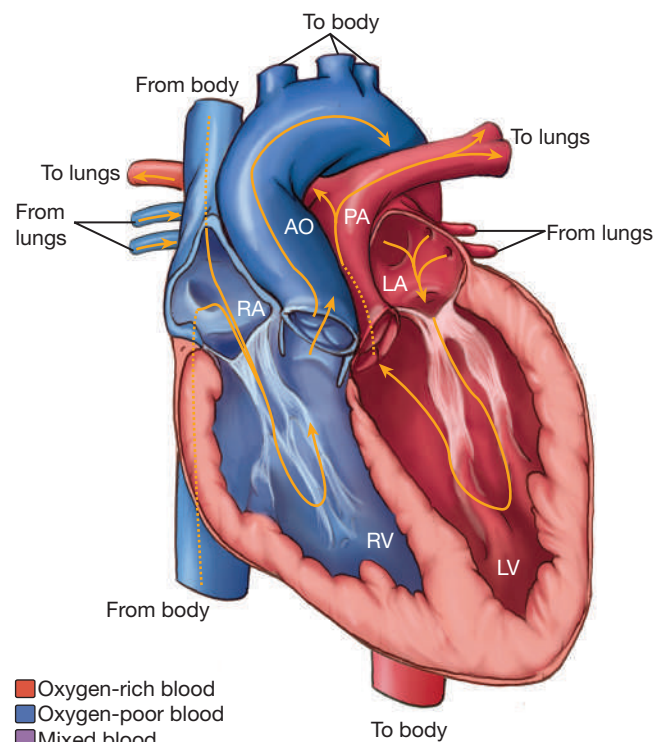


FIGURE 19.11 Transposition of the great vessels.

Health History and Physical Examination

Elicit the health history, noting onset of cyanosis with feeding or crying. Observe the infant for cyanosis while active and at rest. Observe the chest, noting a prominent ventricular impulse. Auscultate the heart, noting a loud second heart sound. A murmur may be heard if the ductus remains open or a septal defect is present. If heart failure is present, note edema, tachypnea, and adventitious lung sounds.

Laboratory and Diagnostic Tests

Echocardiography clearly reveals evidence of the transposition. Cardiac catheterization may be performed to determine whether oxygen saturation levels are low due to the mixing of the blood.

Total Anomalous Pulmonary Venous Connection

TAPVC is a CHD in which the pulmonary veins do not connect normally to the left atrium. Instead, they connect to the right atrium, often by way of the superior vena cava. Relatively rare, it accounts for up to 1.5% of all CHD (Soriano & Fulton, 2022). TAPVC may also be referred to as TAPVR.

PATHOPHYSIOLOGY

Oxygenated blood that would normally enter the left atrium now enters the right atrium and passes to the right ventricle. As a result, the pressure on the right side of the heart increases, leading to hypertrophy. TAPVC is incompatible with life unless there is an associated defect present that allows for shunting of blood from the highly pressured right side of the heart. A patent foramen ovale or an ASD is usually present. Since none of the pulmonary veins connect normally to the left atrium, the only source of blood to the left atrium is blood that is shunted from the right atrium across the defect to the left side of the heart (Fig. 19.12). The highly oxygenated blood from the lungs completely mixes with the poorly oxygenated blood returning from the systemic circulation. This causes an overload of the right atrium and right ventricle. The increased blood volume going into the lungs can lead to pulmonary hypertension and pulmonary edema (Soriano & Fulton, 2022).

NURSING ASSESSMENT

The degree of cyanosis present with TAPVC depends on the extent of the associated defects. For example, if the foramen ovale closes or the ASD is small, significant cyanosis will be present. The physical examination findings will vary depending on the type of TAPVC the infant has, whether obstruction is present, and whether other associated cardiac anomalies are present.

Health History and Physical Examination

Note history of cyanosis, tiring easily, and difficulty feeding. Observe the chest for prominence of the right

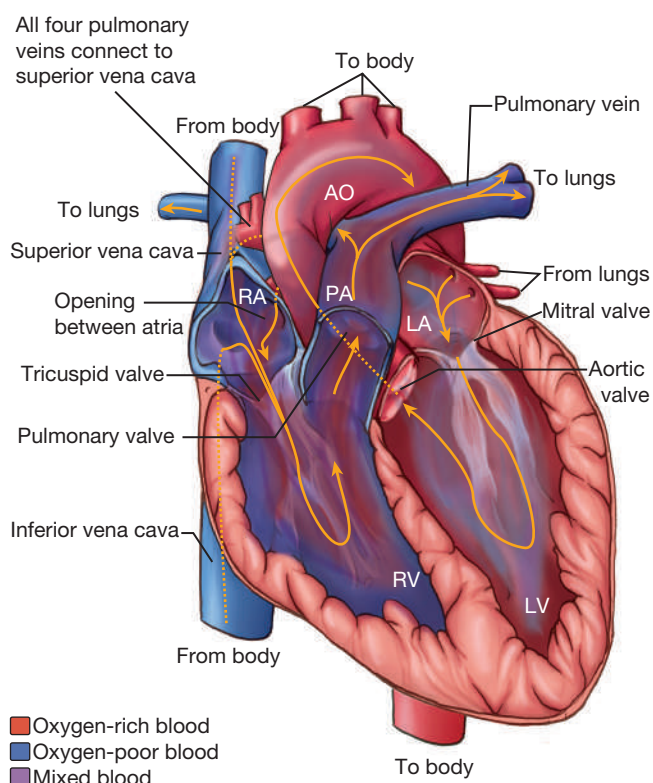


FIGURE 19.12 Total anomalous pulmonary venous connection.

ventricular impulse and retractions with tachypnea. Auscultate the heart, noting fixed splitting of the second heart sound and a murmur. Palpate the abdomen for hepatomegaly.

Laboratory and Diagnostic Tests

An echocardiogram will reveal the abnormal connection of the pulmonary veins, enlargement of the right atrium and right ventricle, and an ASD if present. The chest radiograph will demonstrate an enlarged heart and pulmonary edema. Cardiac catheterization can also be useful to visualize the abnormal connection of the pulmonary veins, particularly if an obstruction is present.

Truncus Arteriosus

Truncus arteriosus is a CHD in which only one major artery leaves the heart and supplies blood to the pulmonary and systemic circulations. It accounts for less than 1% of all CHD cases (Jone et al., 2022; Schneider, 2023). A VSD is almost always present as well.

PATHOPHYSIOLOGY

The one great vessel contains one valve. This valve consists of two to five leaflets and is positioned over both the left and the right ventricles (Fig. 19.13). Due to the location of the valve, blood from the left ventricle mixes with blood from the right ventricle. Pressure

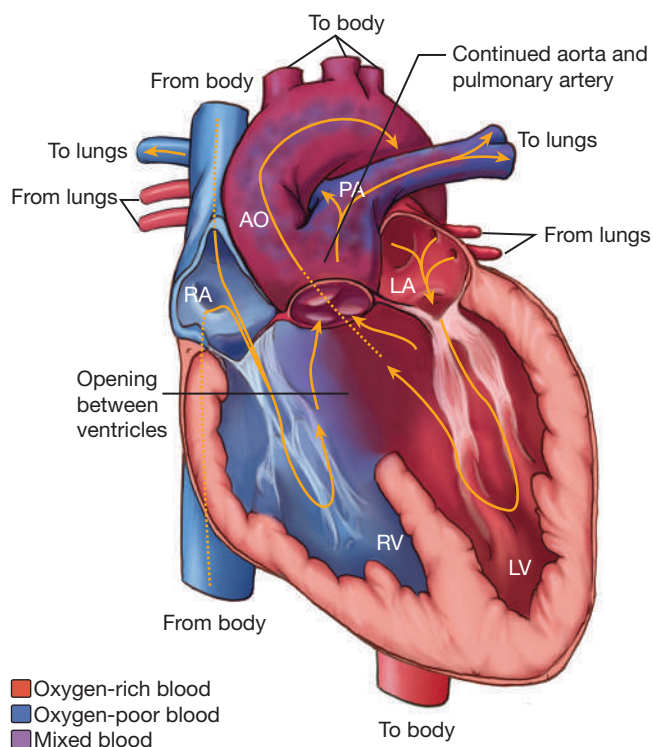


FIGURE 19.13 Truncus arteriosus.

in the pulmonary circulation is typically less than that of the systemic circulation, leading to increased blood flow to the lungs. As a result, systemic blood flow is decreased. Over time, the increased pulmonary blood flow can lead to pulmonary vascular disease (Jone et al., 2022; Schneider, 2023).

NURSING ASSESSMENT

Typically, the infant demonstrates cyanosis in varying degrees, depending on the extent of compromise in the systemic circulation. Obtain an accurate health history and perform a physical examination.

Health History and Physical Examination

Elicit the health history, noting history of cyanosis that increases with periods of activity such as feeding. Also note history of tiring easily, difficulty in feeding, and poor growth. Count the respiratory rate, which may be elevated. Observe for nasal flaring, grunting or noisy breathing, retractions, and restlessness. Auscultate the lungs, noting adventitious breath sounds, and the heart, noting a murmur associated with a VSD.

Laboratory and Diagnostic Tests

An echocardiogram will confirm the presence of truncus arteriosus as the anatomy of the great vessels, the single truncal valve, and the VSD will be seen. On rare occasions, a cardiac catheterization may be done to determine pressures in the pulmonary arteries.

Hypoplastic Left Heart Syndrome

HLHS is a CHD in which all structures on the left side of the heart are severely underdeveloped (Fig. 19.14). The mitral and aortic valves are completely closed or very small. The left ventricle is nonfunctional. Thus, the left side of the heart is completely unable to supply blood to the systemic circulation. HLHS is the fourth most common CHD. It appears to have a multifactorial and autosomal recessive inheritance pattern and occurs in 1.4% to 3.8% of cases of CHD (Jone et al., 2022). The options for treatment include palliative care, cardiac transplantation within the first few weeks of life, or palliative reconstructive surgery consisting of three stages, beginning within days to weeks of birth.

PATHOPHYSIOLOGY

With HLHS, the right side of the heart is the main working part of the heart. Blood returning from the lungs into the left atrium must pass through an ASD to the right side of the heart. The right ventricle must then pump blood to the lungs and to the systemic circulation through the PDA. A few days after birth, when the ductus arteriosus closes, the heart cannot pump blood into the systemic circulation, causing poor perfusion of the vital organs and shock. Death will occur rapidly without intervention (Jone et al., 2022; Schneider, 2023).

NURSING ASSESSMENT

Initially after birth, the newborn may be asymptomatic because the ductus arteriosus is still patent. However, as

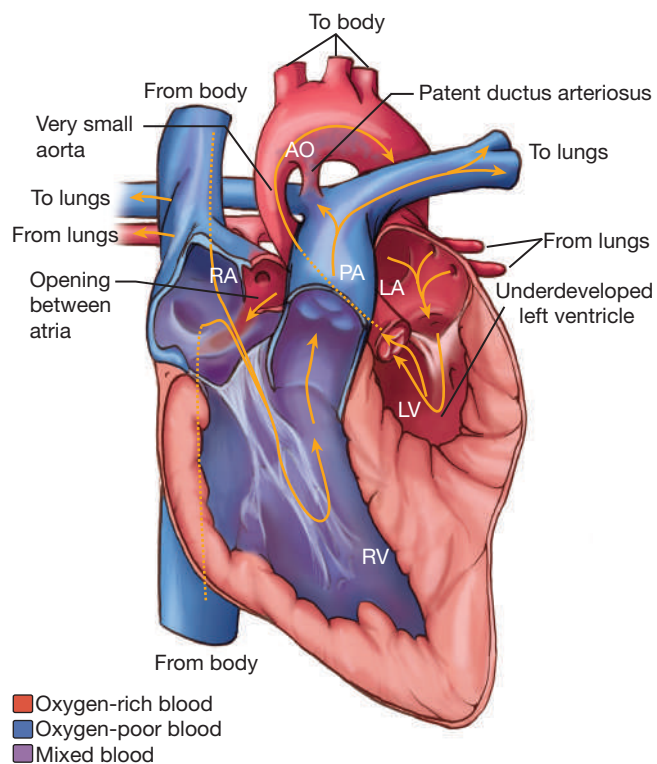


FIGURE 19.14 Hypoplastic left heart syndrome.

the ductus begins to close at a few days of age, the newborn will begin to exhibit cyanosis. Some infants may present with circulatory collapse (shock) and must be resuscitated emergently.

Health History and Physical Examination

Obtain the health history, noting onset of cyanosis. Note poor feeding and history of tiring easily. Evaluate the vital signs, noting tachycardia, tachypnea, and hypothermia. Observe for increased work of breathing and gradually increasing cyanosis. Note pallor of the extremities and decreased oxygen saturation via pulse oximetry. Auscultate the heart and lungs. Note adventitious breath sounds, a gallop rhythm, a single second heart sound, and a soft systolic ejection or holosystolic murmur.

Laboratory and Diagnostic Tests

Prenatally, a fetal echocardiogram can diagnose this syndrome, as can an ultrasound of the pregnant person. After birth, the echocardiogram illustrates the defect.

Nursing Management of the Child With a Congenital Heart Defect

The child with a CHD has multiple needs and requires comprehensive, multidisciplinary care. Nurses play a key role in helping the child and family during this intensely stressful time. Nursing care focuses on improving oxygenation, promoting adequate nutrition, assisting the child and family with coping, providing postoperative nursing care, preventing infection, and providing child and family education. An important component of education involves preparing the child and parents for discharge. In addition to the nursing management presented further on, refer to the “Clinical Judgment and the Nursing Process” section for additional interventions appropriate for the child with CHD. Individualize nursing care specific to the child’s needs.

Improving Oxygenation

Provide frequent ongoing assessment of the child’s cardiopulmonary status as oxygenation status varies due to the hemodynamic changes accompanying the underlying structural defect. Assess airway patency and suction as needed. Position the child in the Fowler or semi-Fowler position to facilitate lung expansion. Monitor vital signs, especially heart and respiratory rates. Monitor the child’s color and oxygen saturation levels closely, using these to guide oxygen administration. Observe for tachypnea and other signs of respiratory distress, such as nasal flaring, grunting, and retractions. Auscultate the lungs for adventitious sounds. Provide humidified supplemental oxygen as ordered, warming it to prevent wide temperature fluctuations. Anticipate the need for assisted ventilation if the child has difficulty maintaining the airway or experiences deterioration in oxygenation capacity. Box 19.1 lists interventions related to relief of hypercyanotic spells.

BOX 19.1 Relieving Hypercyanotic Spells

- Use a calm, comforting approach.
- Place the infant or child in a knee-to-chest position.
- Provide supplemental oxygen.
- Administer morphine sulfate (0.1 mg/kg IV, IM, or SQ).
- Supply IV fluids.
- Administer propranolol (0.1 mg/kg IV).

IM, intramuscularly; IV, intravenously; SQ, subcutaneously.

Data from Doyle, T., & Kavanaugh-McHugh, A. (2023). Management and outcome of tetralogy of Fallot. *UptoDate*. Retrieved January 12, 2024, from <http://www.uptodate.com/contents/management-and-outcome-of-tetralogy-of-fallot>

DOSAGE CALCULATION BOX 19.1

Child’s weight: 12 lb 12 oz

Medication order: digoxin 60 mcg by mouth every 12 hours.

Per the Pediatric Dosage Handbook, the recommended dose is 10–15 mcg/kg/day in two divided doses.

Is the ordered dose safe?

Promoting Adequate Nutrition

Provide nutrition orally, enterally, or parenterally in order to foster growth and development as well as to reduce the risk of infection. The nutritional method will vary depending on the individual child’s energy expenditure associated with increased cardiac and respiratory workloads. In addition, for example, for the newborn or infant, nutrition via breast milk or formula may be provided orally or via gavage feedings. Breastfeeding is usually associated with decreased energy expenditure during the act of feeding, yet some infants in intensive care are not stable enough to breastfeed. Gavage with breast milk is possible, and the use of human milk fortifier (either with breastfeeding or added to the gavage feed) adds additional calories that the infant requires. Formula-fed infants may also require increased-calorie formula, which may be achieved by more concentrated mixing of the formula or through the use of additives such as Polycose or vegetable oil. Consult the nutritionist to determine the individual infant’s caloric needs and prescription of appropriate feeding.

Cutting a larger hole in the nipple or cross-cutting the nipple decreases the work of feeding for some infants. Generally, nipple feedings should be limited to a 20-minute duration, as feeding for longer periods results in excess caloric expenditure. Many infants may feed orally for 20 minutes, receiving the remainder of that feeding via orogastric or nasogastric tube. Offer older children small, frequent feedings to reduce the amount of energy required to feed or eat and to prevent overtiring the child. When needed, administer and monitor total parenteral nutrition as prescribed.

TAKE NOTE!

Breastfeeding a child before and after cardiac surgery may boost the infant's immune system, which can help fight postoperative infection. If breastfeeding is not possible, pumped breast milk may be given via bottle, dropper, or gavage feeding.

Assisting the Child and Family With Coping

Support the family's efforts to cope with the diagnosis of CHD as it can be overwhelming for the child and the parents. The numerous examinations, diagnostic tests, and procedures are sources of stress for the infant or child regardless of age and for the parents. The parents may fear long-term disability or death or may worry that allowing the child to engage in any activity will worsen their status. Thus, the parents may tend to overprotect the child. It is important for the parents to continue parenting the child, even when the child requires extended hospitalizations or intensive care. Explain all that is happening with the child, using language the parents and child can understand. Allow the parents and child to voice their feelings, concerns, or questions. Provide ample time to address these questions and concerns. Encourage the parents and the child, as developmentally appropriate, to participate in the child's care.

If the child is a newborn or infant, encourage attachment and bonding. Emphasize the child's positive attributes, including the normal aspects of the infant. Help the parents to experience the joy of a new infant and see the beauty of the child, no matter how ill the infant is. Urge the parents to touch, stroke, pat, and talk to the infant. Encourage them to hold the infant close, using kangaroo care as appropriate. If the child is older, offer suggestions as to how the parents can meet the child's emotional needs. For example, encourage them to bring a favorite toy or object from home while the child is hospitalized.

Provide developmentally appropriate explanations to the child. Encourage play therapy to help the child understand what is happening.

Preventing Infection

Teach parents proper hand hygiene. Provide appropriate dental care. Make sure the child receives prophylaxis for infective endocarditis as needed. Ensure that children 24 months or younger who are undergoing heart transplantation during respiratory syncytial virus (RSV) season receive appropriate prophylaxis via vaccination with palivizumab (Kimberlin et al., 2021).

Providing Care for the Child Undergoing Cardiac Surgery

Cardiac surgery may be necessary to correct a congenital defect or to provide symptomatic relief. The surgery may

be planned as an elective procedure or done as an emergency. Open heart surgery involves an incision of the heart muscle to repair the internal structures. This may require cardiopulmonary bypass. Closed heart surgery involves structures related to the heart but not the heart muscle itself and may be performed with or without cardiopulmonary bypass.

PROVIDING PREOPERATIVE CARE

Complete the preoperative assessment to provide important baseline information for comparison during the postoperative period. Establish a relationship with the child and parents. Identify problems that may require particular nursing interventions during the postoperative period. Before cardiac surgery, interview the parents and, if age appropriate, the child. Focus the interview on the history of the present illness, cardiac risk factors, the child's present physical and functional status, additional medical problems, current medications and drug allergies, the child's and family's understanding of the illness and planned procedure, and the family support system.

The preoperative physical assessment includes:

- Temperature and weight measurements
- Examination of extremities for peripheral edema, clubbing, and evaluation of peripheral pulses
- Auscultation of the heart (rate, rhythm, heart sounds, murmurs, clicks, and rubs)
- Respiratory assessment, including respiratory rate, work of breathing, and auscultation of the lungs for breath sounds

Obtain any necessary laboratory and diagnostic tests to establish a baseline. In addition, review the results of any tests done previously. Testing may include CBC count, electrolyte levels, clotting studies, urinalysis, cultures of blood and other body secretions, kidney and hepatic function tests, chest radiography, ECG, echocardiogram, and cardiac catheterization.

In most nonemergent cases, preoperative assessment is performed in an outpatient setting, and the child is admitted to the hospital on the day of surgery. Nursing care during this phase focuses on thorough child and parent education. If the surgery is an emergency, teaching must be done quickly, emphasizing the most important elements of the child's care (Beke et al., 2021).

Child and parent education typically includes the following topics:

- Heart anatomy and its function, including what area is involved with the defect that is to be corrected
- Events before surgery, including any testing or preparation such as a skin scrub
- Location of the child after surgery, such as a pediatric intensive care unit, which may include a visit to the unit, if appropriate, and explanation of the sights and sounds that may be present
- Appearance of the child after surgery (equipment or devices used for monitoring, such as oxygen

administration, ECG leads, pulse oximeter, chest tubes, mechanical ventilation, or IV lines)

- Approximate location of the incision and coverage with dressings
- Postoperative activity level, including measures to reduce the risk of complications, such as coughing and deep-breathing exercises, incentive spirometry, early ambulation, and leg exercises
- Nutritional restrictions, such as nothing by mouth for a specified time before surgery and use of IV fluids
- Medications, such as anesthesia, sedation, and analgesics as well as medications the child is taking now that need to be continued or withheld (Beke et al., 2021)

Prepare and educate the child at an age- and developmentally appropriate level. Advise parents to read books with their child about CHD and hospitalization such as:

- *Clifford Visits the Hospital* by N. Bridwell, 2000 (Scholastic Inc.)
- *Franklin Goes to the Hospital* by P. Bourgeois, 2000 (Scholastic Paperbacks)
- *Pump the Bear* by G. O. Whittington, 2000 (Brown Books)
- *Blue Lewis and Sasha the Great* by C. D. Newell, 2005 (Cally Press)
- *Cardiac Kids: A Book for Families Who Have a Child with Heart Disease* by V. Elder, 1994 (Dayton Area Heart and Cancer Association)
- *When Molly Was in the Hospital: A Book for Brothers and Sisters of Hospitalized Children* by D. Duncan, 1994 (Rayve Productions) (siblings)
- *A Night Without Stars* by J. Howe, 1993 (Camelot) (older children)

In addition, parents may order *It's My Heart*, a parent resource book, free of charge from the Children's Heart Foundation via this link: <https://www.childrensheartfoundation.org/about-chds/resources.html>.

Parents may also help their child by buying a small thrift store suitcase, spray painting it, and allowing the child to decorate it with their name, pictures of family, stickers, or favorite story characters. This will be the child's "hospital suitcase" that the child may pack with toys and videos to bring to the hospital. Hospital tours are appropriate for school-age children, and older children and adolescents may benefit from an intensive care unit tour before surgery.

Instruct parents to stop food and liquids at the designated time, depending on the child's age, and to give all medications as directed. Some medications may be withheld before surgery. If the child's nutritional status is poor or questionable, nutritional supplementation may be ordered for a period preoperatively to ensure that the child has the best possible nutritional status before surgery. When it is time for the child to be transported

to the surgical area, allow the parents to accompany the child as far as possible, depending on the institution's policy. Also reinforce with the child that their parents will be present at the bedside when they awaken from surgery (Beke et al., 2021).

PROVIDING POSTOPERATIVE CARE

Postoperative nursing care for the child after cardiac surgery includes the following measures:

- Assess vital signs frequently, as often as every 1 hour, until stable.
- Assess the color of the skin and mucous membranes, check capillary refill, and palpate peripheral pulses.
- Observe cardiac rate and rhythm via electronic monitoring, and auscultate heart rate and rhythm and heart sounds frequently.
- Monitor hemodynamic status via arterial and/or central venous lines (left and right atrial and pulmonary artery pressures, pulmonary artery oxygen saturation).
- Provide site care and tubing changes according to the institution's policy.
- Auscultate lungs for adventitious, diminished, or absent breath sounds.
- Assess oxygen saturation levels via pulse oximetry and arterial blood gases as well as work of breathing and level of consciousness frequently.
- Administer supplemental oxygen as needed.
- Monitor mechanical ventilation and suction as ordered.
- Inspect chest tube functioning, noting amount, color, and character of drainage.
- Inspect the dressing (incision and chest tube) for drainage and intactness. Reinforce or change the dressing as ordered.
- Assess the incision for redness, irritation, drainage, or separation.
- Monitor intake and output hourly.
- Maintain accurate IV infusion rate; restrict fluids as ordered to prevent hypervolemia.
- Assess for changes in level of consciousness. Report restlessness, irritability, or seizures.
- Obtain ordered laboratory tests, such as CBC, coagulation studies, cardiac enzyme levels, and electrolyte levels. Report abnormal results.
- Administer medications, such as digoxin or inotropic or vasopressor agents, as ordered, watching the child closely for possible adverse effects.
- Encourage the child to turn, cough, deep breathe, use the incentive spirometer, and splint the incisional area with pillows.
- Assess the child's pain level and administer analgesics as ordered. Allow time for the child to rest and sleep.
- Assist the child to get out of bed as soon as possible and as ordered.
- Assess daily weights.
- Administer small, frequent feedings or meals when oral intake is allowed.

BOX 19.2 Possible Complications After Cardiac Surgery

- Atelectasis
- Bacterial endocarditis
- Cardiac arrhythmias
- Cardiac tamponade
- Cerebrovascular accident
- Heart failure
- Hemorrhage
- Pleural effusion
- Pneumonia
- Pneumothorax
- Postperfusion syndrome
- Postcardiac surgery syndrome
- Pulmonary edema
- Seizures
- Wound infection

Data from Fleitman, J. (2023). Postoperative complications among patients undergoing cardiac surgery. *UpToDate*. Retrieved January 12, 2024, from <https://www.uptodate.com/contents/postoperative-complications-among-patients-undergoing-cardiac-surgery>

- Position the child in a comfortable position, one that maximizes chest expansion. Change position frequently.
- Assess the child for complications (Box 19.2).
- Provide emotional and physical support to the child and family, making appropriate referrals, such as to social services for assistance.
- Prepare the child and family for discharge (Beke et al., 2021).

TAKE NOTE!

Abrupt cessation of chest tube output accompanied by an increase in heart rate and increased filling pressure (right atrial) may indicate cardiac tamponade (Beke et al., 2021).

Providing Child and Family Education

Provide child and family education throughout the child's stay. Initially, teaching focuses on the underlying defect and measures to treat or control the problem. If the child requires surgery, teaching shifts to preoperative and postoperative events. Emphasize discharge teaching for each admission. Teaching Guidelines 19.2 highlights the major areas to be addressed in child and family education.

ACQUIRED CARDIOVASCULAR DISORDERS

Acquired cardiovascular disorders occur in children because of an underlying cardiovascular problem or may refer to other cardiac disorders that are not congenital. The most common type of acquired cardiovascular

TEACHING GUIDELINES 19.2 Caring for the Child With a Congenital Heart Disease

- Give medications, if ordered, exactly as prescribed.
- Weigh the child at least once a week or as ordered at approximately the same time of the day with the same scale and wearing the same amount of clothing.
- Allow the child to engage in activity as directed. Provide time for the child to rest frequently throughout the day to prevent overexertion.
- Provide a nutritious diet, taking into account any restrictions for fluids or foods.
- Use measures to prevent infection, such as frequent handwashing, prophylactic antibiotics, and skin care.
- Adhere to schedule for follow-up diagnostic tests and procedures.
- Support the child's growth and development needs.
- Use available community support services.
- Notify the health care provider or nurse practitioner if the child has increasing episodes of respiratory distress, cyanosis, or difficulty breathing; fever; increased edema of the hands, feet, or face; decreased urinary output; weight loss or difficulty eating or drinking; increased fatigue or irritability; decreased level of alertness; or vomiting or diarrhea (Gaskin & Kennedy, 2019; Hueckel, 2019).

disorder in children is heart failure. Other acquired disorders include rheumatic fever, cardiomyopathy, infective endocarditis, hyperlipidemia, hypertension, and Kawasaki disease.

Heart Failure

Heart failure refers to a set of clinical signs and symptoms that reflect the heart's inability to pump effectively to provide adequate blood, oxygen, and nutrients to the body organs and tissues (Kusumoto, 2019). Heart failure occurs most often in children with CHD and is the most common reason for admission to the hospital for children with CHD. The estimated number of children experiencing heart failure annually is 12,000 to 25,000 (Singh & Singh, 2022). Heart failure also occurs secondary to other conditions such as myocardial dysfunction following surgical intervention for CHD, cardiomyopathy, myocarditis, fluid volume overload, hypertension, anemia, or sepsis or as a toxic effect of certain chemotherapeutic agents used in the treatment of cancer.

The child experiencing heart failure requires a multidisciplinary approach to care. Collaboration is necessary to achieve improved cardiac function, restored fluid balance, decreased cardiac workload, and improved oxygen delivery to the tissues.

Pathophysiology

Cardiac output is controlled by preload (diastolic volume), afterload (ventricular wall tension), myocardial contractility (inotropic state), and heart rate. Protracted alterations in any of these factors may lead to heart failure. In the event of reduced cardiac output, multiple compensatory mechanisms are activated. When the ventricular contraction is impaired (systolic dysfunction), reduced ejection of blood occurs, and therefore cardiac output is reduced. Diminished ability to receive venous return (diastolic dysfunction) occurs when high venous pressures are required to support ventricular function. As a result of decreased cardiac output, the renin–angiotensin–aldosterone system is activated as a compensatory mechanism. Fluid and sodium retention as well as improved contractility and vasoconstriction then occur. Initially, BP is supported, and organ perfusion is maintained, but increased afterload worsens systolic dysfunction. As the heart chambers dilate, myocardial oxygen consumption increases, and cardiac output is limited by excessive wall stretch. Over time,

the capacity of the heart to respond to these compensatory mechanisms fails, and cardiac output is further decreased (Kusumoto, 2019). Figure 19.15 shows the clinical manifestations that occur related to the mechanisms of heart failure.

Therapeutic Management

Management of heart failure is supportive. Promotion of oxygenation and ventilation is of utmost importance. Digitalis, diuretics, inotropic agents, vasodilators, antiarrhythmics, and antithrombotics have been widely used in children for palliation of symptoms. Many children with heart failure require management in the intensive care unit until they are stabilized. Augmenting nutrition and ensuring adequate rest are also key components of management.

Nursing Assessment

For a full description of the assessment phase of the nursing process, refer to the “Clinical Judgment and the

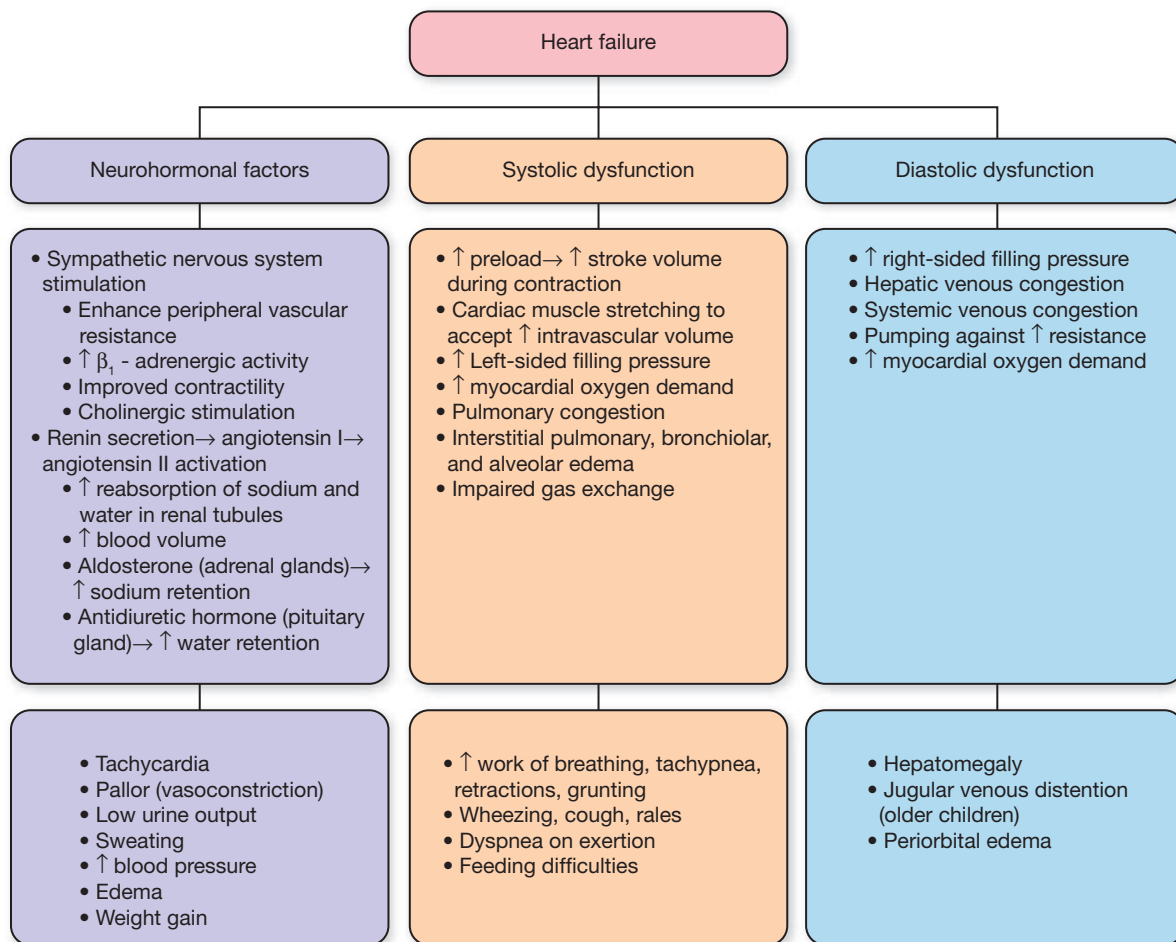


FIGURE 19.15 Pathophysiology of heart failure. (Data from Kusumoto, F. M. [2019]. Cardiovascular disorders: Heart disease. In G. D. Hammer & S. J. McPhee [Eds.], *Pathophysiology of disease: An introduction to clinical medicine* [8th ed.]. McGraw-Hill Education.)

Nursing Process” section earlier in the chapter. Specific assessment findings related to heart failure are discussed further on.

HEALTH HISTORY

When obtaining the health history, elicit a description of the present illness and chief complaint. Common complaints reported during the health history might include:

- Failure to gain weight or rapid weight gain
- Failure to thrive
- Difficulty feeding
- Fatigue
- Dizziness, irritability
- Exercise intolerance
- Shortness of breath
- Sucking and then tiring quickly
- Syncope
- Decreased number of wet diapers

Infants with heart failure often display subtle signs such as difficulty feeding and tiring easily. Pay close attention to reports of these problems from the parents. Also be alert for statements such as “The baby drinks a small amount of breast milk (or formula) and stops but then wants to eat again very soon afterwards;” “The baby seems to perspire a lot during feedings;” or “The baby seems to be more comfortable when he’s sitting up or on my shoulder than when he’s lying flat.” In addition, the parents may report episodes of rapid breathing and grunting.

The child’s current and past medical history also provide additional clues. Question the parents about any history of CHDs and treatments such as surgery to repair the defect. Determine the current medication regimen. Also ask about any recent or past infections, such as streptococcal infections or fever.

PHYSICAL EXAMINATION

Weigh the child and note recent rapid weight gain or lack of weight gain. Obtain the child’s vital signs, noting tachycardia or tachypnea. These findings are often the first indicators of heart failure in an infant or older child. Measure the BP in the upper and lower extremities, comparing the findings for differences. Note decreased BP, which may be due to impaired cardiac muscle function. Inspect the skin color, noting pallor or cyanosis. Also observe for diaphoresis (profuse sweating). Inspect the face, hands, and lower extremities for edema. Observe for increased work of breathing, such as nasal flaring or retractions. Note the presence of a cough, which may be productive with bloody sputum.

Auscultate the apical pulse, noting its location and character. Listen for a murmur, which may suggest a CHD, a gallop rhythm, or an accentuated third heart sound, suggesting sudden ventricular distention. Auscultate the lungs, noting crackles or wheezes suggestive of pulmonary congestion. Palpate the peripheral pulses, noting

weak or thready pulses. Note the temperature and color of the extremities; they may be cool, clammy, and pale. Assess the child’s abdomen, looking for distention indicative of ascites. Gently palpate the abdomen to identify hepatomegaly or splenomegaly.

LABORATORY AND DIAGNOSTIC TESTS

The diagnosis of heart failure is based on the child’s signs and symptoms and is confirmed with several laboratory and diagnostic tests. These include:

- Chest radiograph, revealing an enlarged heart and/or pulmonary edema
- ECG, indicating ventricular hypertrophy
- Echocardiogram, revealing the underlying cause of heart failure, such as a CHD

Other tests may be done to support the diagnosis. For example, the CBC count may show evidence of anemia or infection. Electrolyte levels may reveal hyponatremia secondary to fluid retention and hyperkalemia secondary to tissue destruction or impaired kidney function. Arterial blood gas results may demonstrate respiratory alkalosis in mild heart failure or metabolic acidosis. Tissue hypoxia may be evidenced by increased lactic acid and decreased bicarbonate levels.

Nursing Management

Nursing management of the child with heart failure focuses on promoting oxygenation, supporting cardiac function, providing adequate nutrition, and promoting rest.

PROMOTING OXYGENATION

Position the infant or child in a semi-upright position to decrease work of breathing and lessen pulmonary congestion. Suction as needed. Chest physiotherapy and postural drainage may also be beneficial. Administer supplemental oxygen as ordered and monitor oxygen saturation via pulse oximetry. Oxygen also serves the function of vasodilator and decreases pulmonary vascular resistance. Occasionally, the infant or child with heart failure may require intubation and positive-pressure ventilation to normalize blood gas tension.



CLINICAL REASONING ALERT!

In a child with a large left-to-right shunt, oxygen will decrease pulmonary vascular resistance while increasing the systemic vascular resistance, which leads to increased left-to-right shunting. Monitor the child carefully and use oxygen only as prescribed.

SUPPORTING CARDIAC FUNCTION

Administer digitalis, ACE inhibitors, and diuretics as prescribed. Digoxin therapy begins with a digitalizing dose divided into several doses (oral or IV) over a

24-hour period to reach maximum cardiac effect. During digitalization, monitor the ECG for a prolonged PR interval and decreased ventricular rate. Doses are then administered every 12 hours. Monitor the child for signs of digoxin toxicity. Measure BP before and after administration of ACE inhibitors, holding the dose and notifying the health care provider if the BP falls more than 15 mm Hg. Observe for signs of hypotension such as lightheadedness, dizziness, or fainting. Weigh the child daily to determine fluid loss. Maintain accurate records of intake and output, restricting fluid intake if ordered. Carefully monitor potassium levels, administering potassium supplements if prescribed. Sodium intake is not usually restricted in the child with heart failure.

PROVIDING ADEQUATE NUTRITION

Due to the increased metabolic rate associated with heart failure, the infant may require as much as 150 calories/kg/day. Older children will also require higher caloric intake than typical children. Offer small, frequent feedings if the child can tolerate them. During the acute phase of heart failure, many infants in particular will require continuous or intermittent gavage feeding to maintain or gain weight. Concentrate infant formula to 24 to 28 calories/oz as instructed by the nutritionist.

PROMOTING REST

Minimize metabolic needs to decrease cardiac demand. The infant or older child with heart failure will usually limit activities based on energy level. Ensure adequate time for sleep, and attempt to limit disturbing interventions. Provide age-appropriate activities that can be performed quietly or in bed, such as books, coloring or drawing, and video or board games. The older child or adolescent with significant heart failure may require home schooling. As the child improves, a rehabilitation program may be helpful for maximizing activity within the child's cardiovascular status limits.

Infective Endocarditis

Infective endocarditis is a microbial infection of the endothelial surfaces of the heart's chambers, septum, or valves (most common). Children with CHDs (septum or valve defects) or prosthetic valves are at increased risk for acquiring bacterial endocarditis, which is potentially fatal in these children. Other risk factors for endocarditis include central venous catheters and IV drug use. Infective endocarditis occurs when bacteria or fungi gain access to a damaged epithelium. Turbulence in blood flow associated with narrowed or incompetent valves or with a communication between the systemic and pulmonary circulation leads to damage of the endothelium. Thrombi and platelets then adhere to the endothelium, forming vegetations. When a microbe gains access to the bloodstream, it colonizes the vegetation, using the thrombi as a breeding

ground. Clumps may separate from the vegetative patch and travel to other organs of the body, causing significant damage (septic emboli). Bacteria (particularly alpha-hemolytic streptococcus or *Staphylococcus aureus*) are the most common pathogens responsible for infective endocarditis, and, although rare, *Candida* species may also be found (O'Brien, 2023).

Complete antibiotic or antifungal treatment of the causative organism is necessary, and treatment generally lasts 4 to 6 weeks. Prevention of infective endocarditis in the susceptible child with CHD or a valvular disorder undergoing an invasive procedure is of the utmost importance (O'Brien, 2023).

Nursing Assessment

For a full description of the assessment phase of the nursing process, refer to "Clinical Judgment and the Nursing Process" section. Assessment findings related to endocarditis are discussed further on.

HEALTH HISTORY

Obtain the health history, noting intermittent, unexplained low-grade fever. Document history of fatigue, anorexia, weight loss, or flu-like symptoms (e.g., arthralgia, myalgia, chills, night sweats). Note history of CHD, valve disorder, or heart failure.

PHYSICAL EXAMINATION

Measure the child's temperature, noting low-grade fever. Observe for edema if heart failure is also present. Note petechiae on the palpebral conjunctiva, the oral mucosa, or the extremities. Inspect for signs of extracardiac emboli:

- Roth spots: splinter hemorrhages with pale centers on sclerae, palate, buccal mucosa, chest, fingers, or toes
- Janeway lesions: painless, flat, red or blue hemorrhagic lesions on the palms or the soles
- Osler nodes: small, tender nodules on the pads of the toes or fingers
- Black lines (splinter hemorrhages) under the nails (O'Brien, 2023)

Evaluate the ECG for a prolonged PR interval or dysrhythmias. Auscultate the heart for a new or changing murmur. Auscultate the lungs for adventitious breath sounds. Palpate the abdomen for splenomegaly.

LABORATORY AND DIAGNOSTIC TESTS

Diagnosis is usually based on the clinical presentation. Laboratory tests may reveal the following:

- Blood culture: bacteria or fungus
- CBC: anemia, leukocytosis
- Urinalysis: microscopic hematuria
- Echocardiogram: cardiomegaly, abnormal valve function, area of vegetation

Nursing Management

Nursing management focuses on maintaining IV access for at least 4 weeks to appropriately administer the antibiotic or antifungal course of therapy. Monitor the child's temperature and subsequent blood culture results.

Ideally, infective endocarditis in children should be prevented. Children at increased risk for the development of infective endocarditis include those with:

- Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
- Previous endocarditis
- Unrepaired cyanotic CHD
- Completely repaired CHD with prosthetic material or device within the first 6 months after the procedure
- Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device
- Cardiac transplantation recipients who develop cardiac valve abnormalities (AHA, 2021)

Children at high risk should practice good oral hygiene, including regular toothbrushing and flossing. Instruct parents or the older child to carry emergency medical identification at all times (wallet card is available from the AHA). The card may be presented to any health care provider or nurse practitioner and includes the recommended antibiotic prophylactic regimen (AHA, 2024a). Instruct the parents to notify the primary care provider or cardiologist if the child develops flu-like symptoms or a fever.

High-risk children (as noted previously) who are undergoing dental procedures should receive prophylaxis as recommended by the AHA. Antibiotics typically used for prophylaxis may include ampicillin, amoxicillin, gentamicin, or vancomycin.

Acute Rheumatic Fever

Acute rheumatic fever (ARF) is a delayed sequela of group A streptococcal pharyngeal infection. In the United States, this disease occurs more often in school-age children between 5 and 15 years of age in areas where streptococcal pharyngitis is more prevalent, especially during the colder months. It usually develops 2 to 4 weeks after the initial streptococcal infection. Current understanding of the disease process of ARF is that the child develops an antibody response to surface proteins of the bacteria. The antibodies then cross-react with antigens in cardiac muscle and neuronal and synovial tissues, causing carditis, arthritis, and chorea (involuntary random, jerking movements). ARF affects the joints, central nervous system, skin, and subcutaneous tissue and causes chronic, progressive damage to the heart and valves. Most episodes of ARF resolve, but rheumatic fever may recur with subsequent streptococcal infections (Jone et al., 2022).

Diagnosis of ARF is based on the modified Jones criteria (Box 19.3). Therapeutic management is directed toward managing inflammation and fever, eradicating the bacteria, preventing permanent heart damage, and preventing recurrences. A full 10-day course of penicillin therapy (or equivalent) is used along with corticosteroids and nonsteroidal antiinflammatory drugs. Children without valvular disease will receive continued prophylaxis with monthly intramuscular injections of penicillin G benzathine or daily oral doses of penicillin or erythromycin following the initial illness to prevent a new streptococcal infection and recurrent ARF. Prophylaxis is usually continued until age 21 years (Jone et al., 2022).

Nursing Assessment

Elicit a description of the present illness and chief complaint, noting fever and joint pain. Explore the child's recent medical history for risk factors, such as documented streptococcal infection or sore throat within the past 2 to 3 weeks, or for history of ARF. Observe the child for Sydenham chorea, a movement disorder of the face and upper extremities. Inspect the skin for evidence of the classic rash, erythema marginatum, a maculopapular red rash with central clearing and elevated edges. Auscultate the heart, noting a murmur. Palpate the surfaces of the wrist, elbows, and knees for firm, painless, subcutaneous nodules. Note prolonged PR interval on the ECG. Throat culture will provide definitive diagnosis of current streptococcal infection, while streptococcal antibody tests may yield evidence of recent infection. Echocardiogram is required to determine if carditis is present.

Nursing Management

Nursing management of the child with ARF focuses on ensuring compliance with the acute course of antibiotics

BOX 19.3 Modified Jones Criteria

Diagnosis of ARF requires the presence of either two major criteria or one major plus two minor criteria.

Major Criteria

- Carditis
- Migratory polyarthritis
- Subcutaneous nodules
- Erythema marginatum
- Sydenham chorea

Minor Criteria

- Polyarthralgia
- Elevated ESR or CRP
- Prolonged PR interval (unless carditis is a major criterion)

ARF, acute rheumatic fever; CRP, C-reactive protein; ESR, erythrocyte sedimentation
Data from Jone, P.-N., Kim, J. S., Burkett, D., Jacobsen, R., & VonAlvensleben, J. (2022). Cardiovascular diseases. In M. Bunik, W. W. Hay, M. J. Levin, & M. J. Abzug (Eds.), *Current diagnosis and treatment: Pediatrics* (26th ed.). McGraw-Hill Education.

as well as prophylaxis following initial recovery from ARF. Allow the child to verbalize the frustration they may be feeling in relation to chorea symptoms. Offer support for dealing with the abnormal movements. Educate the child and others that the sudden jerky movements of chorea will eventually disappear, although they may last as long as several months. Some children may require a neuroleptic agent such as haloperidol (Haldol) for management of chorea. Administer corticosteroids or nonsteroidal antiinflammatory agents for control of joint pain and swelling.

Cardiomyopathy

Cardiomyopathy is a condition in which the myocardium cannot contract properly. The incidence of cardiomyopathy among children is increasing; it occurs at a rate of one per 100,000 (AHA, 2024b). Cardiomyopathy may occur in children with genetic disorders or CHDs, as a result of an inflammatory or infectious process or hypertension, or after cardiac transplantation or surgery, but most commonly, it is idiopathic. Cardiomyopathy occurs predominantly in clusters in infancy and adolescence. Three types of cardiomyopathy exist—restrictive, dilated, and hypertrophic. Restrictive cardiomyopathy is rare in children and results in atrial relaxation. Dilated cardiomyopathy is the most common type in childhood and may result in heart failure (it may be their presentation) because of ventricular dilation with decreased contractility (Jones et al., 2022). There is also some familial tendency toward dilated cardiomyopathy (Cooper, 2022). Hypertrophic cardiomyopathy is more common in adolescence and results in hypertrophy of the heart muscle, particularly the left ventricle, affecting the heart's ability to fill. About two-thirds of all cases of hypertrophic cardiomyopathy are familial, with some inherited in an autosomal dominant fashion (Cooper, 2022).

There is no cure for cardiomyopathy, meaning that currently, heart muscle function cannot be restored. Therapeutic management is directed toward improving heart function and BP. Mechanical ventilation and vasoactive medications are needed in many children. ACE inhibitors, beta-blockers, or calcium channel blockers may be used. Pacemakers or surgery may be helpful in some children. For children in whom medical management is unsuccessful, heart transplantation is the only viable long-term treatment option (Jones et al., 2022).

Nursing Assessment

Explore the health history for risk factors such as:

- CHD, cardiac transplantation, or surgery
- Duchenne or Becker muscular dystrophy
- History of myocarditis, HIV infection, or Kawasaki disease

- Hypertension
- Drugs, alcohol, or radiation exposure
- Connective tissue, autoimmune, or endocrine disease
- Maternal diabetes
- Familial history of sudden death

Inquire about a history of respiratory distress, fatigue, poor growth (dilated), chest pain, dizziness, or syncope (hypertrophic). Observe the child for extremity edema and abdominal distention. Note increased work of breathing. Auscultate the heart, noting tachycardia and irregular rhythm. Evaluate heart rhythm via ECG, noting dysrhythmias or indications of left ventricular hypertrophy.

Chest radiography may reveal cardiomegaly or congested lungs. Echocardiogram demonstrates increased heart size, poor contractility, decreased ejection fraction, or asymmetric septal hypertrophy. Cardiac catheterization is usually performed to aid in the diagnosis.

Nursing Management

Many children with cardiomyopathy require intensive care initially. Monitor for complications such as blood clots or arrhythmias, which could lead to cardiac arrest. Refer to the previous section on heart failure for nursing interventions related to heart failure, which may be present with dilated cardiomyopathy. Administer vasoactive and other medications as prescribed, monitoring the child closely for response to these therapies as well as for complications. Support the child in choosing activities that fit within the prescribed restrictions. Provide extensive emotional support to the child and family, who may experience significant stress as they realize the severity of this illness.

Hypertension

Hypertension has seen a rise in prevalence among children and adolescents. It has been found to be independently associated with body mass index and waist circumference. Childhood or adolescent hypertension often leads to long-term health consequences such as cardiovascular disease and left ventricular hypertrophy (Mattoo, 2023). In children, acceptable BP values are based on sex, age, and height. For children age 1 to 13 years, stage 1 hypertension is defined as BP persistently greater than or equal to the 95th percentile for sex, age, and height or less than the 95th percentile plus 12 mm Hg (whichever is lower). Stage 2 hypertension in children is identified as BP greater than or equal to the 95th percentile plus 12 mm Hg or 140/90, whichever is lower. For adolescents 13 years and older, stage 1 hypertension is defined as BP 130/80 to 139/89, while stage 2 hypertension is identified as BP greater than or equal to 140/90. The term “elevated blood pressure” refers to BP

that is persistently between the 90th and 95th percentiles or 130/80 (whichever is lower) in children up to age 13 years. In adolescents 13 years of age or older, elevated BP refers to systolic blood pressure of 120 to 129, with diastolic BP less than 80. BP is considered normal when the systolic and diastolic values are less than the 90th percentile for sex, age, and height in the child 1 to 13 years of age or less than 120/80 in the adolescent 13 years and older (Flynn et al., 2017).

Childhood hypertension may be further defined as primary or secondary. Primary hypertension in children is found more commonly in non-Hispanic African Americans and children with overweight or obesity (Mattoo, 2023). Secondary hypertension in children most frequently occurs with an underlying medical problem such as kidney or cardiac disease (Mattoo, 2023). Mild to moderate hypertension in childhood is usually asymptomatic and is usually determined only upon BP screening during a well-child visit or during follow-up for known risk factors. Refer to Box 9.5 in Chapter 9 for a synopsis of childhood hypertension guidelines.

It is important to screen for and treat prehypertension and hypertension in children and adolescents, as they are more likely to experience hypertension as adults progressing to further cardiovascular disease (Mattoo, 2021). Therapeutic management depends on the extent of the hypertension and the length of time it has existed. Weight reduction, appropriate diet (including sodium restriction in some children), and increased physical activity are important components of management of prehypertensive and asymptomatic hypertensive children. Some children are candidates for and require antihypertensive medications or diuretics (Mattoo, 2021).

Pathophysiology

The balance between cardiac output and vascular resistance determines the BP. An increase in either of these variables, in the absence of a compensatory decrease in the other, increases the mean BP. Factors regulating cardiac output and vascular resistance include changes in electrolyte balance, particularly sodium, calcium, and potassium.

Nursing Assessment

Nursing assessment consists of the health history, physical examination, and laboratory and diagnostic tests.

HEALTH HISTORY

Elicit the health history, determining the presence of risk factors for hypertension such as:

- Family history
- Obesity
- Hyperlipidemia

- Kidney disease (including frequent urinary tract infections)
- Systemic lupus erythematosus
- CHD
- Neurofibromatosis, Turner syndrome, and other genetic disorders
- Prematurity
- Prolonged neonatal ventilation
- Umbilical artery catheterization
- Diabetes mellitus
- Increased intracranial pressure
- Malignancy
- Solid organ transplant
- Medications known to raise BP

Signs and symptoms reported during the health history might include:

- Growth delays (with certain chronic medical conditions)
- Obesity
- Signs and symptoms seen particularly in older children
- Headache
- Subtle behavioral or school performance changes
- Fatigue
- Blurred vision
- Nosebleed
- Bell palsy

PHYSICAL EXAMINATION

Determine the child's weight and height/length. Plot these growth parameters on the sex-appropriate chart for the child's age. Note the percentile for height/length, as it will be used to determine the BP percentile (see Appendix B, "Blood Pressure Charts for Children and Adolescents"). Measure the BP in all four extremities (to rule out coarctation of the aorta). Ensure that the child is relaxed and sitting or reclined. Refer to Chapter 10 for specific information related to accurate BP measurement in children.

Inspect the skin for:

- Acne, hirsutism, or striae (associated with anabolic steroid use)
- Café-au-lait spots (associated with neurofibromatosis)
- Malar rash (associated with lupus)
- Pallor, diaphoresis, or flushing (associated with pheochromocytoma)

Observe the extremities for edema (kidney disease) or joint swelling (lupus). Inspect the chest for apical heave (ventricular hypertrophy) or wide-spaced nipples (Turner syndrome). Auscultate heart sounds, noting tachycardia (associated with primary hypertension) or murmur (associated with coarctation of the aorta). Palpate the abdomen for a mass or enlarged kidney.

LABORATORY AND DIAGNOSTIC TESTING

Although diagnosis of hypertension is based on BP measurements, additional laboratory or diagnostic tests may

be used to evaluate the underlying cause of secondary hypertension, including:

- Urinalysis, blood urea nitrogen, and serum creatinine: may determine the presence of kidney disease
- Renal ultrasound or angiography: may reveal kidney or genitourinary tract abnormalities
- Echocardiogram: may show left ventricular hypertrophy
- Lipid profile: determines the presence of hyperlipidemia

Nursing Management

Salt restriction and potassium or calcium supplements have not been scientifically shown to decrease BP in children. However, children with obesity may benefit from salt restriction, as those children seem to be sensitive to salt intake. Assist the child and family to develop a plan for weight reduction if the child has overweight or obesity. Encourage the child and family to control portion sizes, decrease the intake of sugary beverages and snacks, eat more fresh fruits and vegetables, and eat a healthy breakfast. Consult the nutritionist for additional assistance with meal planning. To increase physical activity, encourage the child to find a sport or type of exercise in which they are interested. Aerobic activities involving running, walking, or cycling are particularly helpful. When a child requires antihypertensive therapy, teach the child and family how to administer the medication. Caution the parents about side effects related to antihypertensives. Teach the parent to measure the child's BP as determined by the health care provider or nurse practitioner, as well as to keep appointments for BP follow-up.

Kawasaki Disease

Kawasaki disease is an acute systemic vasculitis occurring mostly in children 6 months to 5 years of age. It is

the leading cause of acquired heart disease among children and in the United States, occurs more than 19 times per year per 100,000 children (Lo et al., 2025). Although Kawasaki disease affects all ethnic groups, it occurs more frequently in those of Asian or Pacific descent. It is a self-limited syndrome but can cause cardiovascular complications, such as coronary artery aneurysm and cardiomyopathy (Lo et al., 2025).

Therapeutic management of acute Kawasaki disease focuses on reducing inflammation in the walls of the coronary arteries and preventing coronary thrombosis. Chronic management of children developing aneurysms during the initial phase is directed toward preventing myocardial ischemia. In the acute phase, high-dose aspirin in four divided doses daily and a single infusion of intravenous immunoglobulin (IVIG) are used (Lo et al., 2025). See Evidence-Based Practice 19.1.

Pathophysiology

Although the etiology is still unknown, current thought is that some infectious organism (as yet unidentified) causes disease in genetically susceptible people. Kawasaki disease appears to be an autoimmune response mediated by cytokine-induced endothelial cell surface antigens that leads to vasculitis in the medium-size arteries, including the coronary arteries. Neutrophils, mononuclear cells, T lymphocytes, and immunoglobulin A–producing plasma cells infiltrate the vessels. Then, elastin and collagen fibers fragment, and the structural integrity of the vessel wall are impaired. Generalized systemic vasculitis occurs in the blood vessels throughout the body due to the inflammation and edema and can lead to coronary dilation or aneurysm. Some children never develop coronary artery changes, while others develop an aneurysm in either the acute phase or as a long-term sequela (Lo et al., 2025).

EVIDENCE-BASED PRACTICE 19.1

Treating Kawasaki Disease With Intravenous Immunoglobulin (IVIG)

STUDY

Coronary artery abnormalities remain the most serious complication of the acute vasculitis occurring in Kawasaki disease. Historically, IVIG of varying doses and other medications such as aspirin and corticosteroids have been used to reduce the risk of coronary artery anomaly (CAA) development. The study explored the use of IVIG in the acute phase. In their review, the authors included 31 studies with 4,609 participants.

Findings

It was determined that high-dose IVIG provided during the acute phase probably reduced the risk of development of CAA as compared to the use of moderate- or low-dose IVIG, aspirin, or corticosteroids.

The occurrence of adverse effects was low for all treatment regimens.

Nursing Implications

The study results are consistent with the current recommendations for treatment of Kawasaki disease. Teach families that administration of IVIG is safe and that it is used to reduce the risk of CAA development. Refer to Chapter 25 for additional information related to IVIG administration.

Based on Broderick, C., Kobayashi, S., Suto, M., Ito, S., & Kobayashi, T. (2023). Intravenous immunoglobulin for the treatment of Kawasaki disease. *Cochrane Database of Systematic Reviews*, 1, CD014884. <https://doi.org/10.1002/14651858.CD014884.pub2>

Nursing Assessment

Nursing assessment consists of determining the health history, physical examination, and laboratory and diagnostic testing.

HEALTH HISTORY

Elicit the health history, noting any:

- Fever
- Chills
- Headache
- Malaise
- Extreme irritability
- Vomiting
- Diarrhea
- Abdominal pain
- Joint pain

Of note is a history of high fever (39.9°C [103.8°F]) of at least 5 days' duration that is unresponsive to antibiotics.

PHYSICAL EXAMINATION

Observe for significant bilateral conjunctivitis without exudate. Inspect the mouth and throat for dry, fissured lips; strawberry (cracked and reddened) tongue; and pharyngeal and oral mucosa erythema. Note hyperdynamic precordium. Evaluate the skin for:

- Diffuse, erythematous, polymorphous rash
- Edema of the hands and feet
- Erythema and painful induration of the palms and soles
- Desquamation (peeling) of the perineal region, fingers, and toes, extending to the palms and soles
- Possible jaundice

Palpate the neck for cervical lymphadenopathy (usually unilateral) and the joints for tenderness. Palpate the abdomen for liver enlargement. Auscultate the heart, noting tachycardia, gallop, or murmur.

LABORATORY AND DIAGNOSTIC TESTING

The CBC may reveal mild to moderate anemia, an elevated white blood cell count during the acute phase, and significant thrombocytosis (elevated platelet count [500,000 to 1 million]) in the later phase. The erythrocyte sedimentation rate (ESR) and the C-reactive protein (CRP) level are elevated. Echocardiogram is performed as soon as possible after the diagnosis is confirmed to provide a baseline of a healthy heart or to evaluate for coronary artery involvement. Echocardiograms may be repeated during the illness and as part of long-term follow-up. Occasionally, cardiac involvement warrants cardiac catheterization.

Nursing Management

In addition to the administration of aspirin and immunoglobulin, nursing management of the child with Kawasaki disease focuses on monitoring cardiac status, promoting comfort, and providing family education.

MONITORING CARDIAC STATUS

Administer IV and oral fluids as ordered, evaluating intake and output carefully. Prepare the child for the echocardiogram. Assess frequently for signs of developing heart failure such as tachycardia, gallop, decreased urine output, or respiratory distress. Evaluate quality and strength of pulses. Provide cardiac monitoring as ordered, reporting arrhythmias.

PROMOTING COMFORT

Provide acetaminophen for fever management, and apply cool cloths as tolerated. Keep the environment quiet, and cluster nursing care activities to decrease stimulation and hence irritability. Teach parents that irritability is a prominent feature of Kawasaki disease, and support their efforts to console the child. Apply petrolatum jelly or another lubricating ointment to the lips. Encourage the older child to suck on ice chips; the younger child may suck on a cool, moist washcloth. Popsicles are also soothing. Provide comfortable positioning, particularly if the child has joint pain or arthritis.

PROVIDING CHILD AND FAMILY EDUCATION

Teach parents to continue to monitor the child's temperature after discharge until the child has been afebrile for several days. Children with prolonged or recurrent fever may require a second dose of IVIG. Inform parents that irritability may last for up to 2 months after initial diagnosis with Kawasaki disease. Report any toxic effects of aspirin therapy, such as headache, confusion, dizziness, or tinnitus to the health care provider or nurse practitioner. It is important to avoid nonsteroidal antiinflammatory agents while aspirin therapy is ongoing. For children with continued arthritis (which resolves in several weeks), range-of-motion exercises with a morning bath may help to decrease stiffness. Instruct parents to avoid measles and varicella vaccination for 11 months after high-dose IVIG administration. It is critical that the family comply with regularly scheduled cardiology follow-up appointments to determine development or progression of coronary artery ectasia or aneurysm. If the child has severe cardiac involvement, teach the parents about infant and/or child cardiopulmonary resuscitation before discharge from the hospital.

Dyslipidemia

Dyslipidemia refers to high levels of lipids (fats/cholesterol) in the blood. High lipid levels are a risk factor for the development of atherosclerosis, which can result in coronary artery disease, a serious cardiovascular disorder occurring in adults. Children with high lipid levels, although remaining asymptomatic, are likely to have high levels as adults, which increases their risk for coronary artery disease. Therefore, detection, screening, and early intervention are important, especially if there is a family tendency toward heart disease (de Ferranti & Newburger, 2023b).

Pathophysiology

Cholesterol is a building block for hormones and cell membranes. It occurs naturally in foods derived from animals such as eggs, dairy products, meat, poultry, and seafood. Cholesterol is also manufactured in the body. Together, cholesterol and triglycerides are known as lipids. Very low-density lipoprotein (VLDL) is a lipoprotein composed mainly of triglycerides with only small amounts of cholesterol, phospholipid, and protein. VLDLs are easily converted to low-density lipoproteins (LDLs). Cholesterol is expressed in terms of LDL cholesterol or high-density lipoprotein (HDL) cholesterol. LDLs contain relatively more cholesterol and triglycerides than they do protein. HDLs contain about 50% protein, with the rest being cholesterol, triglyceride, and phospholipid. High levels of cholesterol and triglycerides place a person at risk for atherosclerosis. Elevated VLDL and LDL levels and decreased HDL levels produce a particular increase in the risk for atherosclerosis (de Ferranti & Newburger, 2023b).

Therapeutic Management

Screening children for hyperlipidemia is of prime importance for early detection, intervention, and subsequent prevention of adult atherosclerosis. The American Academy of Pediatrics recommends universal screening for dyslipidemia between 9 and 11 years of age and again between 18 and 21 years of age (Hagan et al., 2017). Performing a risk assessment screening at 24 months and at 4, 6, 8, and 12 through 17 years of age is also recommended. Selectively screening children at high risk for hyperlipidemia can reduce their lifelong risk of coronary artery disease. The risk assessment focuses on the child's family history. Screen if parents, grandparents, aunts and uncles, or siblings, have or have had documented:

- Coronary atherosclerosis
- Myocardial infarction
- Angina pectoris
- Peripheral vascular disease
- Cerebrovascular disease/stroke
- Coronary artery bypass graft/stent/angioplasty at less than 55 years of age in males and less than 65 years in females
- Sudden cardiac death
- Blood cholesterol level of 240 mg/dL or higher

The child should be screened at the health care provider's discretion if the parental history is unobtainable, the child has diabetes or hypertension, or the child has any lifestyle risk factors (cigarette smoking, obesity, sedentary lifestyle, or high-fat dietary intake) (de Ferranti & Newburger, 2023b).

All children should eat a diet with the appropriate amount of fats (see the section on nursing management further on) and should participate in physical activity. When diet and exercise are not enough to lower cholesterol to appropriate levels, medications such as statins may be used (Jone et al., 2022).

Nursing Assessment

Elicit the health history, noting risk factors such as family history of hyperlipidemia, early heart disease, hypertension, diabetes or other endocrine abnormality, cerebral vascular accident, or sudden death. Note prior lipid levels if available. Measure the child's height and weight, plotting them on standardized growth charts. Note if the child has overweight or obesity, as these are risk factors associated with hyperlipidemia. Typically, there are no other particular physical findings associated with hyperlipidemia. Table 19.3 gives details about the interpretation of cholesterol levels.

Nursing Management

Instruct families that the child must fast for 12 hours before lipid screening (initially and on follow-up samples). Dietary management is the first step in the prevention and management of hyperlipidemia in children older than 2 years. The diet should consist primarily of fruits, vegetables, low-fat dairy products, whole grains, beans, lean meat, poultry, and fish. As in adults, fat should account for no more than 30% of daily caloric intake. Fat intake may vary over a period of days, as many young children are picky eaters. Limit saturated fats by choosing lean meats; removing skin from poultry before cooking; and avoiding palm, palm kernel, and coconut oils as well as hydrogenated fats. Teach families to read nutrition labels to determine the content of the food. Limit intake of processed or refined foods as well as high-sugar drinks; these products provide minimal nutrition and significant

TABLE 19.3 • Interpretation of Cholesterol Levels for Children and Adolescents

Total Cholesterol (mg/dL)	Interpretation	LDL (mg/dL)	Interpretation	HDL (mg/dL)	Interpretation
<170	Desirable	<110	Optimal	35	Desirable
170–199	Borderline			110–129	Borderline
≥200	High			>130	High

HDL, high-density lipoprotein; LDL, low-density lipoprotein

Data from de Ferranti, S. D., & Newburger, J. W. (2023b). Dyslipidemia in children and adolescents: Definition, screening, and diagnosis. *UpToDate*. Retrieved January 12, 2024, from <https://www.uptodate.com/contents/dyslipidemia-in-children-and-adolescents-definition-screening-and-diagnosis>

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calories. Children 5 to 10 years of age need vigorous play or physical activity for 1 hour per day, three times per week, while children older than 10 years of age should participate in vigorous activity 60 minutes daily (de Ferranti & Newburger, 2023a). Refer parents to “Healthy Habits for Healthy Kids—A Nutrition and Activity Guide for Parents” published by the American Dietetic Association and available at http://www.clocc.net/wp-content/uploads/Healthy_Habits_Healthy_Kids.pdf.

If medications are required, teach the child and family about the dose, administration, and possible adverse effects. Assist the family to develop a medication-dosing plan that is compatible with school and work schedules to increase compliance.

HEART TRANSPLANTATION

Heart transplantation is indicated in children with inoperable CHD or with end-stage heart disease related to cardiomyopathy or palliated CHD. Worldwide, 600 to 700 children receive a heart transplant each year (Bock & Chinnock, 2022). The 5-year survival rate is greater than 70%, and 20-year survival has been achieved in some instances (Bock & Chinnock, 2022).

A comprehensive evaluation is performed to determine whether the child is a candidate for heart transplant. The evaluation includes:

- Chest radiograph, ECG, echocardiogram, exercise stress test, cardiac catheterization, and pulmonary function tests
- CBC with differential, prothrombin and partial thromboplastin time, serum chemistries and electrolytes, blood urea nitrogen, and creatinine
- Urinalysis and urine creatinine clearance
- Blood, throat, urine, stool, and sputum cultures for bacteria, viruses, fungi, and parasites
- Epstein–Barr virus, cytomegalovirus, varicella, herpes, hepatitis, and HIV titers
- Human leukocyte antigen (HLA) typing and panel reactive antibody typing and titer
- Computed tomography or MRI scan and electroencephalogram
- Consults with neurology, psychology, genetics, social work, nutritionist, physical and occupational therapy, and financial coordinator or case manager (Bock & Chinnock, 2022)

Children with irreversible lung, liver, kidney, or central nervous system disease; recent malignancy (past 5 years); or chronic viral infection may be excluded as candidates.

Once candidacy is determined, the transplant center registers the child as a potential recipient with the United Network for Organ Sharing (UNOS). Blood type, body size, length of time on the waiting list, and medical urgency are used to evaluate compatibility. Children awaiting transplantation may need continuous or intermittent hospitalization. Coordination of organ procurement and the transplantation procedure is essential.

Surgical Procedure and Postoperative Therapeutic Management

Most transplantation procedures are **orthotopic**, which means that the recipient’s heart is removed and the donor heart is implanted in its place in the normal anatomic position. Cardiopulmonary bypass and hypothermia are used to maintain circulation, protect the brain, and oxygenate the recipient during the procedure. Postoperatively, the child may have near-normal heart function and capacity for exercise and may be able to return to school.

Immunosuppressive therapy is necessary for the rest of the child’s life to avoid rejection of the transplanted heart. Usually, a three-drug regimen is used that includes calcineurin inhibitors (cyclosporine, tacrolimus), cell toxins (mycophenolate mofetil, azathioprine), and corticosteroids. The cardiologist and transplant surgeon provide ongoing follow-up. Complications of heart transplantation include infection, pulmonary hypertension, arrhythmia, heart failure, hypertension, kidney dysfunction, and organ rejection. Neoplasm may occur as a result of chronic immunosuppression.

Nursing Management

Preoperative nursing care for the child undergoing a heart transplant is similar for children undergoing other types of heart surgery. In addition, the nurse should assist with the comprehensive pretransplant evaluation. Care for the child in the posttransplant period is intense and complex. Evaluate the family’s ability to perform the tasks that will be necessary. Teach families about the evaluation and transplantation process, as well as the waiting period. In the immediate preoperative period, perform a thorough history and physical examination, and obtain last-minute blood work. Provide preoperative teaching similar to other cardiac surgeries. Older children, adolescents, and parents may enjoy the book *Future Conditional* by J. Hatton (1996, Yorkshire Art Circus), which was written by one of the first heart transplant survivors.

Postoperatively, provide frequent assessments and routine care for children who have had cardiac surgery. In addition, monitor the child closely for infection or signs of rejection. Acute rejection may be indicated by low-grade fever, fatigue, tachycardia, nausea, vomiting, abdominal pain, and decreased activity tolerance, although some children will be asymptomatic. Maintain strict handwashing techniques and isolate the child from other children with infections. Although live vaccines are contraindicated in children with immunosuppression, inactivated vaccines should be given as recommended (CDC, 2020). Teach children and families that the child may return to school and usual activities about 3 months after the transplant. Provide emotional support to the child related to body image changes such as hair growth, gum hyperplasia, weight gain, moon facies, acne, and rashes that occur due to long-term immunosuppressive therapy.

KEY CONCEPTS

- At birth, when the umbilical cord is cut and the neonate's first breath occurs, the ductus venosus closes with the foramen ovale, and the ductus arteriosus closes shortly thereafter. Pulmonary vascular resistance decreases, and systemic vascular resistance increases.
- The infant's heart rate averages 120 to 130 bpm and decreases throughout childhood, reaching the adult rate in adolescence. Conversely, the infant's and child's BP is significantly lower than the adult's, increasing as the child ages.
- Check the infant's apical pulse prior to digoxin administration, and hold the dose if the heart rate is less than 90.
- Poor weight gain, failure to thrive, and increased fatigability commonly occur with congestive heart failure.
- Clubbing of the fingernails occurs because of chronic hypoxia in the child with severe CHD.
- Children with cardiac conditions resulting in cyanosis often have baseline oxygen saturations that are relatively low, because of the mixing of oxygenated with deoxygenated blood.
- Document the presence of a murmur by grading its intensity (I through IV), describing where it occurs within the cardiac cycle, and noting the location where the murmur is best heard.
- CHD should be suspected in the cyanotic newborn who does not improve with oxygen administration.
- Cardiac catheterization postprocedure care focuses on evaluation of the child's vital signs and condition of the pressure dressing, as well as assessment of the distal pulses bilaterally for presence and quality.
- Congenital heart disorders resulting in decreased pulmonary blood flow (tetralogy of Fallot, tricuspid atresia) result in cyanosis.
- Disorders with increased pulmonary blood flow (PDA, ASD, and VSD) may result in pulmonary edema if the defect is severe.
- A decrease in the lower extremity pulses or BP as compared with the upper extremities may be indicative of coarctation of the aorta.
- It is important to remain calm when an infant or child demonstrates a hypercyanotic spell. Place the child in a knee-chest position, administer oxygen and/or morphine or propranolol, and supply IV fluids.
- Children with certain CHDs and/or heart failure require additional calories to display adequate growth.
- Children with hypertrophic cardiomyopathy, certain CHDs, valve dysfunction, or prosthetic valves require prophylaxis for infective endocarditis when undergoing procedures or invasive dental work.
- Hypertension in the child or adolescent often leads to long-term health consequences such as cardiovascular disease and left ventricular hypertrophy.
- Kawasaki disease may result in severe cardiac sequelae, so these children need ongoing cardiac follow-up to screen for development of problems.

- It is important to screen for hyperlipidemia in high-risk children.
- Abrupt cessation of chest tube output, accompanied by an increase in the heart rate and increased filling pressure, may indicate cardiac tamponade.

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DEVELOPING CLINICAL JUDGMENT

PRACTICING FOR NCLEX

- The nurse is caring for a 5-year-old child with a congenital heart anomaly causing chronic cyanosis. When performing the history and physical examination, what is the nurse least likely to assess?
 - Obesity from overeating
 - Clubbing of the nail beds
 - Squatting during play activities
 - Exercise intolerance
- A 2-day-old infant was just diagnosed with aortic stenosis. What is the most likely nursing assessment finding?
 - Gallop and rales
 - Blood pressure discrepancies in the extremities
 - Right ventricular hypertrophy on ECG
 - Heart murmur
- Sam, age 11, has a diagnosis of rheumatic fever and has missed school for a week. What is the most likely cause of this problem?
 - Previous streptococcal throat infection
 - History of open heart surgery at 5 years of age
 - Playing too much soccer and not getting enough rest
 - Exposure to a sibling with pneumonia
- The nurse is caring for a child after a cardiac catheterization. What is the nursing priority?
 - Allow early ambulation to encourage activity participation.
 - Check pulses above the catheter insertion site for strength and quality.
 - Assess extremity distal to the insertion site for temperature and color.
 - Change the dressing to evaluate the site for infection.
- While assessing a 4-month-old infant, the nurse notes that the baby experiences a hypercyanotic spell. What is the priority nursing action?
 - Provide supplemental oxygen by face mask.
 - Administer a dose of IV morphine sulfate.
 - Begin cardiopulmonary resuscitation.
 - Place the infant in a knee-to-chest position.
- The nurse is providing discharge instructions to the parent of a 2-month-old infant who has been prescribed digoxin to be administered every 12 hours orally. Which instructions should the nurse include in the discharge instructions? Select three items.
 - Notify the health care provider or nurse practitioner if more than two consecutive doses are missed.
 - Mix the medication with a small amount of formula or breast milk.

- If the infant demonstrates poor feeding or vomiting, notify the health care provider or nurse practitioner.
 - If the child vomits immediately after administration, repeat the dose.
 - As soon as it is noted that a dose has been missed, give the medication.
 - Always give the medication at regular intervals.
- An adolescent patient was admitted with a sore throat, a red rash on the trunk, swollen and painful joints, and aimless movements of the extremities. The diagnosis of ARF is made.

Vital Signs

Time	Temperature	Apical Heart Rate	Respiratory Rate	Blood Pressure
0800	38.0°C	94	22	110/80
1200	37.1°C	142	24	120/84

What should the nurse do first?

- Administer prescribed acetaminophen.
- Apply moisturizer to the adolescent's rash.
- Notify the health care provider or nurse practitioner of the vital signs change.
- Splint the affected joints to relieve pain.

DOSAGE CALCULATION QUESTION

The nurse is caring for an infant with a VSD who has heart failure. The infant weighs 11 lb. The medication order reads: spironolactone 5 mg PO every 12 hours. Spironolactone is provided by the pharmacy in a solution of 2.5 mg/1 mL. How many milliliters will the nurse administer? Round to the nearest whole number.

CRITICAL THINKING EXERCISES

- A baby was born at 26 weeks' gestation to 15-year-old parents with substance use disorder. The infant weighed 1.5 kg at birth and was diagnosed with AV canal defect and Down syndrome. Discuss some of the major issues in planning for care. Include a care plan and a list of teaching needs for the family.
- A 4-year-old has parents with less than a high school education, and the child has Medicaid coverage. Another child is 7 years old and has parents with advanced degrees and private insurance coverage. Both children need a heart transplant, and a heart is available that is a good match for both children. Discuss some of the issues involved in deciding which child should receive the heart.
- A 13-year-old was diagnosed with hypertension more than 2 years ago. He is nonadherent to his antihypertensive medication regimen.

He is 5 ft tall and weighs 170 lb. His favorite activity is video games. Develop a teaching plan for this adolescent, providing creative approaches at the appropriate developmental level.

STUDY ACTIVITIES

1. Teach a class of sixth graders about healthy activities to prevent high cholesterol levels, hypertension, and heart disease. Use visual materials.
2. Spend the day with a nurse practitioner in the pediatric cardiology clinic. Report to the clinical group your observations about the children's quality of life, growth, and development.
3. Observe in the pediatric cardiothoracic intensive care unit or telemetry unit. Note the different cardiac rhythms displayed by children with a variety of cardiovascular disorders.