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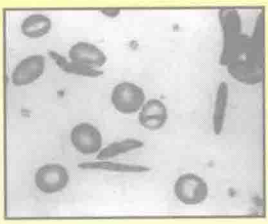
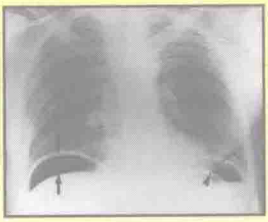
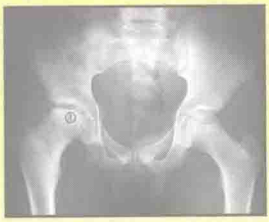
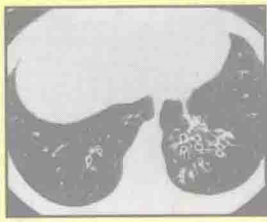

## PEDIATRICS

*Classic Clinical Cases for USMLE Step 2 and Clerkship Review*


VIKAS BHUSHAN, MD
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HOANG NGUYEN
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S2S
STUDENT TO STUDENT

# UNDERGROUND
# CLINICAL VIGNETTES

## PEDIATRICS

*Classic Clinical Cases for*
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# Preface

This series was developed to address the nearly universal presence of clinical vignette questions on the USMLE Step 2. It is designed to supplement and complement *First Aid for the USMLE Step 2* (Appleton & Lange). Bidirectional cross-linking to appropriate High-Yield Facts in the second edition of *First Aid for the USMLE Step 2* has been implemented.

Each book uses a series of approximately 50 **"supra-prototypical" cases as a way to condense testable facts and associations.** The clinical vignettes in this series are designed to incorporate as many testable facts as possible into a cohesive and memorable clinical picture. The vignettes represent composites drawn from general and specialty textbooks, reference books, thousands of USMLE-style questions and the personal experience of the authors and reviewers. Additionally, we present "Associated Diseases" as a way to teach the most critical facts about a larger number of diseases that do not justify an entire case. **The "Associated Diseases" list is NOT complete and does not represent differential diagnoses.**

Although each case tends to present all the signs, symptoms, and diagnostic findings for a particular illness, **patients generally will not present with such a "complete" picture either clinically or on the Step 2 exam.** Cases are not meant to simulate a potential real patient or an exam vignette. All the **boldfaced "buzzwords" are for learning purposes** and are not necessarily expected to be found in any one patient with the disease. **Similarly, the images for each case are for learning purposes only, were derived from a variety of textbooks, and may not match the clinical vignette in all respects.** Images are labeled [A]–[D] and represent 1–4 images of varying sizes, with locations corresponding to a left-to-right, top-to-bottom lettering system.

Definitions of selected important terms are placed within the vignettes in (= SMALL CAPS) in parentheses. Other parenthetical remarks often refer to the pathophysiology or mechanism of disease. The format should also help students learn to present cases succinctly during oral "bullet" presentations on clinical rotations. The cases are meant to be read as a condensed review, not as a primary reference.

The information provided in this book has been prepared with a great deal of thought and careful research. This book should not, however, be considered your sole source of information. Corrections, suggestions, and submissions of new cases are encouraged and will be acknowledged and incorporated in future editions.

# Abbreviations

ABGs - arterial blood gases
ACTH - adrenocorticotropic hormone
ADA - adenosine deaminase
AIDS - acquired immunodeficiency syndrome
ALL - acute lymphocytic leukemia
ALT - alanine transaminase
AMP - adenosine monophosphate
ANA - antinuclear antibody
Angio - angiography
AP - anteroposterior
ARDS - adult respiratory distress syndrome
ASD - atrial septal defect
ASO - anti-streptolysin O
AST - aspartate transaminase
AZT - zidovudine
BCG - bacille Calmette–Guérin
BP - blood pressure
BUN - blood urea nitrogen
CALLA - common acute lymphocytic leukemia antigen
CBC - complete blood count
CDC - Centers for Disease Control
CF - cystic fibrosis
CFTR - cystic fibrosis transmembrane conductance regulator
CHF - congestive heart failure
CK - creatine kinase
CNS - central nervous system
COPD - chronic obstructive pulmonary disease
CRP - C-reactive protein
CSF - cerebrospinal fluid
CT - computed tomography
CXR - chest x-ray
DIC - disseminated intravascular coagulation
DMD - Duchenne's muscular dystrophy
DTP - diphtheria/tetanus/pertussis
DTRs - deep tendon reflexes
DVT - deep venous thrombosis
EBV - Epstein–Barr virus
ECG - electrocardiography
Echo - echocardiography
ECMO - extracorporeal membrane oxygenation
EEG - electroencephalography
ELISA - enzyme-linked immunosorbent assay
EMG - electromyography
ESR - erythrocyte sedimentation rate
FEV - forced expiratory volume
FTA-ABS - fluorescent treponemal antibody absorption
FVC - forced vital capacity
GA - gestational age

# Abbreviations - continued

G6PD - glucose-6-phosphate dehydrogenase
GI - gastrointestinal
Hb - hemoglobin
HGPRT - hypoxanthine-guanine phosphoribosyl transferase
HIV - human immunodeficiency virus
HLA - human leukocyte antigen
HPI - history of present illness
HR - heart rate
ID/CC - identification and chief complaint
IFA - immunofluorescent antibody
Ig - immunoglobulin
IM - intramuscular
INH - isoniazid
IVC - inferior vena cava
JRA - juvenile rheumatoid arthritis
JVP - jugular venous pressure
KUB - kidneys/ureter/bladder
LDH - lactate dehydrogenase
LFTs - liver function tests
LP - lumbar puncture
L/S - lecithin-to-sphingomyelin (ratio)
LV - left ventricular
LVH - left ventricular hypertrophy
Lytes - electrolytes
MI - myocardial infarction
MMR - measles/mumps/rubella
MR - magnetic resonance (imaging)
NG - nasogastric
NPO - nil per os (nothing by mouth)
NSAID - nonsteroidal anti-inflammatory drug
Nuc - nuclear medicine
PBS - peripheral blood smear
PCR - polymerase chain reaction
PDA - patent ductus arteriosus
PE - physical exam
PFTs - pulmonary function tests
PMI - point of maximal intensity
PPD - purified protein derivative
PT - prothrombin time
PTT - partial thromboplastin time
RBC - red blood cell
RF - rheumatoid factor
RPR - rapid plasma reagin
RR - respiratory rate
RSV - respiratory syncytial virus
RV - right ventricular
RVH - right ventricular hypertrophy
SBFT - small bowel follow-through

# Abbreviations - continued

SIDS - sudden infant death syndrome
TMP-SMX - trimethoprim-sulfamethoxazole
TSH - thyroid-stimulating hormone
UA - urinalysis
UGI - upper GI
URI - upper respiratory infection
US - ultrasound
VMA - vanillylmandelic acid
VS - vital signs
VSD - ventricular septal defect
vWF - von Willebrand factor
WBC - white blood cell
XR - x-ray

**ID/CC** A 2-year-old **female** presents with **poor feeding** and **difficulty breathing.**

**HPI** She was born in a small town in the Rocky Mountains (**high altitude** predisposes) and was delivered at 28 weeks' gestation (more common in **preterm** infants). On directed questioning, her mother recalls that she had a transitory skin rash during the first trimester of her pregnancy (**rubella** predisposes).

**PE** VS: tachycardia; tachypnea. PE: **no cyanosis; bounding arterial pulses; wide pulse pressure;** hyperdynamic LV impulse displaced laterally; **continuous "machinery murmur"** noted at second and third left intercostal space lateral to sternal border.

**Labs** ECG: left axis deviation; LVH.

**Imaging** CXR: **increased pulmonary vascular markings;** enlarged left ventricle, left atrium, pulmonary arteries, and ascending aorta; the ductus arteriosus may show calcification. Echo: enlarged left atrium and ventricle. Angio: **increased oxygen saturation in the pulmonary artery** (diagnostic).

**Pathogenesis** Failed closure of fetal communication between the pulmonary artery and aorta; commonly associated with maternal rubella and coxsackievirus infection, premature birth, and respiratory distress syndrome. The ductus normally closes as a result of increased oxygen tension during the first 48 hours of life (may take up to three weeks). The persistent communication between the descending aorta and pulmonary artery near the left subclavian artery increases pulmonary blood flow in systole and diastole, causing pulmonary congestion and LV overload.

**Epidemiology** **Twice as common in females;** more common in infants born at high altitudes and in premature infants.

**Management** In the presence of respiratory distress syndrome, **treat heart failure** (diuretics, digitalis) and anemia. **Indomethacin,** a prostaglandin E1 inhibitor, may stimulate ductus closure. **Surgery** consists of simple ligation (preferred), clipping, or division and may be considered in the absence of pulmonary hypertension. Administer **prophylactic antibiotics** with dental and

surgical procedures.

**Complications**

If left untreated, there is a high risk of **left heart failure** (most common), **infective endocarditis, endarteritis,** and **Eisenmenger's syndrome** (symptomatic pulmonary hypertension resulting from high pulmonary vascular flow; eventually leads to the development of a right-to-left shunt; manifests as right heart failure and cyanosis).

**Associated Diseases**

◘ **Atrial Septal Defect** The most common congenital heart disease in adults; acyanotic left-to-right shunt of left atrium into right atrium; presents with shortness of breath and systolic ejection murmur; fixed, wide split of S2; ECG shows right axis deviation; CXR shows increased pulmonary vascularity; treat by surgical repair; complications include paradoxic emboli, pulmonary hypertension, and Eisenmenger's syndrome (due to reversal of shunt).

◘ **Ventricular Septal Defect** An acyanotic cardiac malformation leading to left-to-right shunt; presents with dyspnea, parasternal heave, and pansystolic murmur; ECG reveals biventricular hypertrophy; CXR shows cardiomegaly; treat by surgical repair.

**ID/CC** A 9-year-old girl presents with **shortness of breath** (= DYSPNEA) mostly while running or playing, coupled with lightheadedness and **easy fatigability** (due to decreased cardiac output).

**HPI** Yesterday she complained of **severe chest pain** while skipping rope. She has no history of allergies, surgery, trauma, transfusions, hospitalizations, or major illnesses. Her vaccinations are up to date. The mother states that the child was born with **congenital rubella** (predisposing factor).

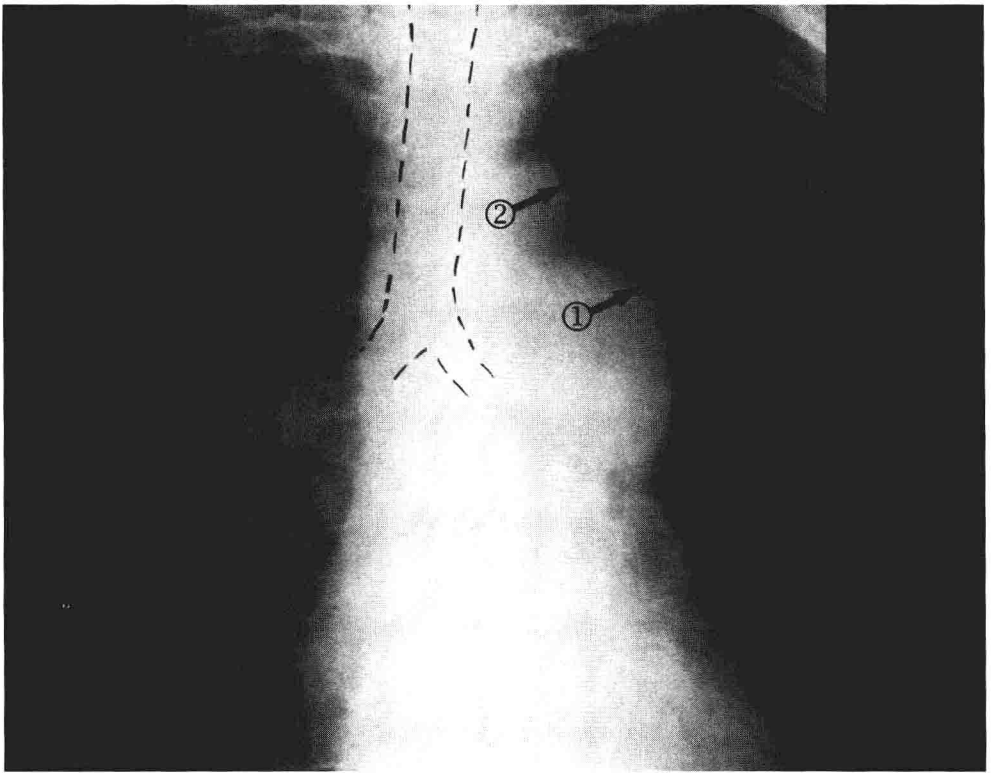
**PE** VS: normal. PE: **raised JVP** with prominent "a" wave; presystolic liver pulsation (increased venous pressure); palpable **RV heave;** crescendo-decrescendo (diamond-shaped) **systolic ejection murmur** preceded by click in left second interspace (pulmonary area) radiating to neck; **soft P2 and widely split S2.**

**Labs** CBC/Lytes: normal. ECG: right axis deviation; RV enlargement.

**Imaging** [A] CXR: **poststenotic dilated pulmonary artery** (1); note the relative size of the aortic knob (2). Other findings include **diminished pulmonary vascular markings.** Echo: RV enlargement; dome-shaped valve. Angio: diagnostic; RVH with a transpulmonary gradient.

**Pathogenesis** A **cyanotic** congenital heart disease that is **idiopathic,** although some viral infections have been implicated (congenital rubella is a predisposing factor). In the **neonatal** period, patients may present with **cyanosis** (right-to-left shunt through patent foramen ovale); mild disease may be asymptomatic. In moderate to severe disease there may be exertional dyspnea, hypoxic spells, squatting episodes (more typical of tetralogy of Fallot), and even ischemic chest pain. It may be isolated but is more commonly associated with a patent foramen ovale or with other cardiac defects, such as VSD, ASD, and PDA. May be **valvular, infundibular,** or combined. Associated with Noonan's syndrome and malignant intestinal carcinoid.

**Epidemiology** Fifty percent of deaths occur within the first year of life unless a compensatory shunt (e.g., VSD, ASD, PDA)




persists.

**Management**

**Prostaglandin E1** keeps the ductus arteriosus patent until surgery in neonates. **Balloon valvuloplasty** (mainly for isolated pulmonary stenosis) or surgical repair is required if the transpulmonary valve gradient exceeds 50 mmHg. **Emergent surgery** is indicated in acute right heart failure. RVH usually resolves after corrective surgery. Patients should be given antibiotic prophylaxis for **infective endocarditis** before dental and surgical procedures.

**Complications**

Complications include **cardiac failure** (most common), sudden death (most frequently in infancy), low cardiac output, growth retardation, hypoxic spells, and arrhythmias. Postoperative complications include recurrence (mainly if surgery was done early) and pulmonary insufficiency.

**Associated Diseases**

■ **Coarctation of the Aorta** Congenital stenosis of the aorta usually distal to the left subclavian artery; increased incidence in patients with Turner's syndrome; presents with weak and delayed femoral pulses, upper extremity hypertension, and systolic murmur heard loudest over the back; often asymptomatic but can cause claudication