Theodore J. Saclarides
Jonathan A. Myers
Keith W. Millikan
Editors

Common Surgical Diseases

An Algorithmic Approach to Problem Solving

Third Edition

Springer

Common Surgical Diseases

Third Edition

Common Surgical Diseases

An Algorithmic Approach to Problem Solving

Third Edition

Edited by

Theodore J. Saclarides
Professor of Surgery, Head
Division of Colon and Rectal Surgery
Loyola University Medical Center
Maywood, IL, USA

Jonathan A. Myers
Department of General Surgery
Rush University Medical Center
Chicago, IL, USA

Keith W. Millikan
Department of General Surgery
Rush University Medical Center
Chicago, IL, USA

Springer

Editors
Theodore J. Saclarides
Division of Colon and Rectal Surgery
Loyola University Medical Center
Maywood, IL, USA

Keith W. Millikan
Department of General Surgery
Rush University Medical Center
Chicago, IL, USA

Jonathan A. Myers
Department of General Surgery
Rush University Medical Center
Chicago, IL, USA

ISBN 978-1-4939-1564-4 ISBN 978-1-4939-1565-1 (eBook)
DOI 10.1007/978-1-4939-1565-1
Springer New York Heidelberg Dordrecht London

Library of Congress Control Number: 2014948462

Printed on acid-free paper

Springer is part of Springer Science+Business Media (www.springer.com)

I dedicate this work to Elena for her support and understanding and my children Kathryn (Kashi), Eddie (mi hijo), Stephanie (Weph), Constantine (Deno), Alexandra (Zaz), and Theodora (Dofro). My unconditional love goes to my granddaughter Salome who, like the myrrh bearing women, inspires an understanding of the totality of our existence, a faith that should remain steadfast, and a life that is eternal.

Theodore J. Saclarides

This book is dedicated to my wife, Beth, for her love and support and to our children, Jack and Megan, for their spirit and enthusiasm.

Jonathan A. Myers

To my daughter Samantha, who I hope can use this book as an aid for caring for patients as she pursues a career in medicine.

Keith W. Millikan

Preface

Written by leaders in the field, *Common Surgical Diseases: An Algorithmic Approach to Problem Solving* provides surgical residents and house staff with a current, concise and algorithmic approach to frequently encountered clinical challenges. More than 90 chapters detail every common surgical disease in the form of a succinct text coupled with a step-by-step algorithm. Each chapter walks the reader through the evaluation, diagnosis, treatment, and follow-up of the most common surgical problems.

Thoroughly updated and revised, the third edition includes new chapters on DVT prophylaxis, access for hemodialysis, adrenal incidentaloma, esophageal cancer, pancreatic cancer, genetic predisposition to colorectal cancer and breast cancer, abdominal wall defects, hyperglycemia, necrotizing soft tissue infections, and SIRS/sepsis.

Following the success of the first two editions, this revision provides an excellent quick reference and is an absolute essential for practicing surgeons, surgical house staff, and for medical students in their surgical clerkship.

Theodore J. Saclarides
Maywood, IL, USA
Jonathan A. Myers
Chicago, IL, USA
Keith W. Millikan
Chicago, IL, USA

Contents

Contributors

Chetan Aher, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Rosalinda Alvarado, M.D.
Department of General Surgery, Johns Hopkins Medicine – Suburban Hospital, Bethesda, MD, USA

Andrew T. Arndt, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA; Department of Cardiothoracic Surgery, Yale University, New Haven, CT, USA

Bernadette Aulivola, M.D.
Department of Surgery, Division of Vascular Surgery and Endovascular Therapy, Loyola University Medical Center, Maywood, IL, USA

Paul R. Balash, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

David Baldwin Jr., M.D.
Endocrinology, Rush University Medical Center, Chicago, IL, USA

Rana M. Ballo, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Jonas S. Benson, M.D.
Department of Urology, UroPartners, Wheaton, IL, USA

Elizabeth Berger, M.D.
General Surgery, Loyola University Medical Center, Maywood, IL, USA

Kristin Bevil
Loyola University Stritch School of Medicine, Maywood, IL, USA

Steven Bines, M.D.
Rush University Medical Center, Chicago, IL, USA

Jaqueline Blank
Loyola University Stritch School of Medicine, Maywood, IL, USA

Alexi Bloom
Loyola University Stritch School of Medicine, Maywood, IL, USA

Lisa N. Boggio, M.D.
Rush Hemophilia and Thrombophilia Center, Department of Medicine and Pediatrics, Hematology, Rush University Medical Center, Chicago, IL, USA

Julia Boll, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Leon Boudourakis, M.D.
Department of Surgery, The Mount Sinai Medical Center, New York, NY, USA

Marc I. Brand, M.D.
Department of General Surgery, Section of Colorectal Surgery, Rush University Medical Center, Chicago, IL, USA

Joseph Broucek, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Sherry Cavanagh, M.D.
Department of Cardiovascular/Thoracic Surgery, Rush University Medical Center, Chicago, IL, USA

Edie Y. Chan, M.D., FACS
Department of General Surgery, Section of Transplantation, Rush University Medical Center, Chicago, IL, USA

John D. Christein, M.D.
Department of Surgery, Section of Gastrointestinal Surgery, University of Alabama-Birmingham, Birmingham, AL, USA

Jamie J. Coleman, M.D.
Indiana University Department of Surgery, Indiana University Methodist Hospital, Indianapolis, IN, USA

Kyle G. Cologne, M.D.
Division of Colorectal Surgery, Keck School of Medicine, University of Southern California, Los Angeles, CA, USA

Christopher L. Coogan, M.D.
Department of Urology, Rush University Medical Center, Chicago, IL, USA

Marie Crandall, M.D., MPH, FACS
Department of Surgery, Northwestern University Feinberg School of Medicine, Chicago, IL, USA

John Cull, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Shaun Daly, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Steven A. De Jong, M.D., FACS, FACE
Department of Surgery, Division of General Surgery, Perioperative Services, Loyola University Medical Center, Maywood, IL, USA

Daniel J. Deziel, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Michael W. Dingeldein, M.D.
Department of Pediatric Surgery, The Children's Hospital at the University of Oklahoma Medical Center, Oklahoma City, OK, USA

Nadav Dujovny, M.D.
Colon and Rectal Surgery, Spectrum Health, The Ferguson Clinic, Grand Rapids, MI, USA

Erin Farlow, M.D.
Department of Vascular Surgery, Indiana University Health Methodist Hospital, Indianapolis, IN, USA

Leandro Feo, M.D.
The Ferguson Clinic/MMPC, Grand Rapids, MI, USA

Jonathan M. Frank, M.D.
Department of Orthopedic Surgery, Rush University Medical Center, Chicago, IL, USA

Loretto Ann Glynn, M.D.
Department of Pediatric Surgery, Lurie Children's Hospital at Cadence Health, Winfield, IL, USA

Adam B. Goldin, M.D., MPH
Department of Pediatric General and Thoracic Surgery, Seattle Children's Hospital, Seattle, WA, USA

Mehra Golshan, M.D.
Breast Surgical Services, Department of Surgery, Brigham and Women's Hospital and Dana Farber/Brigham and Women's Cancer Center, Boston, MA, USA

Kristin Gross, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Alicia Growney, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Alfred Guirguis, DO, MPH
Department of Obstetrics and Gynecology, Rush University Medical Center, Chicago, IL, USA

Elizabeth C. Gwinn, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Erica Hammes, MMS, PA-C
Department of Kidney Transplant, Advocate Christ Medical Center, Oak Lawn, IL, USA

Dana M. Hayden, M.D., MPH
Department of General Surgery, Division of Colon and Rectal Surgery, Loyola University Medical Center, Maywood, IL, USA

Katherine Heiden, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Michael P. Hoeh, M.D.
Department of Urology, Rush University Medical Center, Chicago, IL, USA

Edward F. Hollinger, M.D., Ph.D.
Department of General Surgery, Section of Abdominal Transplant, Rush University Medical Center, Chicago, IL, USA

Hadyn Hollister, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Edward Hong, M.D.
Department of Thoracic Surgery, Rush University Medical Center, Chicago, IL, USA

Keith C. Hood, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Elizabeth A. Hooper, M.D.
Department of General Surgery, Mercy Clinic General Surgery – Mercy Plaza, Oklahoma City, OK, USA

Tasha M. Hughes, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Matthew J. Hyser, M.D., FACS
Department of Surgery, Metropolitan Group Hospitals, St. Francis Hospital, Surgical Associates S.C., Evanston, IL, USA

Chad E. Jacobs, M.D.
Department of Cardiovascular/Thoracic Surgery, Rush University Medical Center, Chicago, IL, USA

Jennifer Jolley, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Nikolaos D. Karagiorgos, M.D.
Department of Surgery, Metropolitan Group Hospital General Surgery, University of Illinois, Advocate Illinois Masonic Medical Center, Chicago, IL, USA

Anna B. Katz, M.D.
Department of Breast Surgery, Advocate Medical Group, Libertyville, IL, USA

Clifford J. Kavinsky, M.D., Ph.D.
Section of Cardiology, Rush University Medical Center, Chicago, IL, USA

Abid Khan, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Anthony W. Kim, M.D.
Section of Thoracic Surgery, Department of Surgery, Yale New Haven Hospital, New Haven, CT, USA

Yoojin Kim, M.D.
Department of Endocrincology, Rush University Medical Center, Chicago, IL, USA

Matthew Klairmont, M.D.
Rush Medical College, Chicago, IL, USA

Ryan C. Knoper, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Chung-Kay Koh, M.D.
Department of Internal Medicine, Rush University Medical Center, Chicago, IL, USA

Katherine Kopkash, M.D.
Department of Surgery, Rush University Medical Center, Chicago, IL, USA

Vikram D. Krishnamurthy, M.D.
Department of Surgery, Rush University Medical Center, Chicago, IL, USA

Stephen M. Larsen, M.D.
Department of Urology, Rush University Medical Center, Chicago, IL, USA

Kalyan C. Latchamsetty, M.D.
Department of Urology, Rush University Medical Center, Chicago, IL, USA

Marissa Le, M.D.
Department of Otolaryngology, Rush University Medical Center, Chicago, IL, USA

Cynthia L. Leaphart, M.D.
Transplantation Division of Transplant Surgery, Mayo Clinic Florida, Jacksonville, FL, USA

Benjamin B. Lind, M.D.
Division of Vascular Surgery, NorthShore University Health System, Skokle, IL, USA

Phillip S. LoSavio, M.D.
Department of Otolaryngology – Head and Neck Surgery, Rush University Medical Center, Chicago, IL, USA

Li Shien Low, M.D.
Section of Cardiology, Rush University Medical Center, Chicago, IL, USA

James L. Lubawski Jr., M.D.
Department of Cardiothoracic Surgery, Cedars-Sinair Medical Center, Los Angeles, CA, USA

Minh B. Luu, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Andrea Madrigrano, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Mike Malinowski, M.D.
Department of Vascular Surgery, Loyola University Medical Center, Maywood, IL, USA

Patri Marconi, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Joseph M. Maurice, M.D.
Section of Gynecology, Minimally Invasive Gynecologic Surgery, Department of Obstetrics and Gynecology, Rush University Medical Center, Chicago, IL, USA

Jarod P. McAteer, M.D., MPH
Department of Surgery, University of Washington, Seattle, WA, USA

John Christopher McAuliffe, M.D., Ph.D.
Department of Surgery, University of Alabama – Birmingham Hospital, Birmingham, AL, USA

Amina Merchant, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Marc G. Mesleh, M.D.
Department of General Surgery, Advocate Christ Hospital, Oak Lawn, IL, USA

Thomas Messer, M.D.
Department of Surgical Critical Care, Stroger Hospital of Cook County, Chicago, IL, USA

Keith W. Millikan, M.D.
Department of Surgery, Rush University Medical Center, Chicago, IL, USA

Deepak Mital, M.D.
Transplant Surgery, Advocate Christ Medical Center, Oak Lawn, IL, USA

Tricia A. Moo-Young, M.D.
Surgical Oncology, NorthShore Medical Group, Highland Park, IL, USA

Maria C. Mora Pinzon, M.D.
Division of Colon and Rectal Surgery, Loyola University Medical Center, Maywood, IL, USA

Jonathan A. Myers, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Oyedolamu K. Olaitan, M.D.
Section of Transplantation, Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Purvi P. Patel, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Dana K. Perry, M.D.
Department of Transplant Surgery, Mayo Clinic Florida, Jacksonville, FL, USA

Lindsay Petersen, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Srikumar Pillai, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Andrew M. Popoff, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Shobha L. Rao, M.D.
Department of Internal Medicine, Rush University Medical Center, Chicago, IL, USA

Morgan K. Richards, M.D.
Department of General Surgery, University of Washington School of Medicine, Seattle, WA, USA

Harry Richter, M.D.
Department of Surgery, Stroger Cook County Hospital, Chicago, IL, USA

Jennifer Rickard, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Daniel Rinewalt, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Bruce A. Rosenzweig, M.D.
Section of Urogynecology, Department of Obstetrics and Gynecology, Rush University Medical Center, Chicago, IL, USA

Theodore J. Saclarides, M.D.
Section of Colon and Rectal Surgery, Department of Surgery, Loyola University Medical Center, Maywood, IL, USA

Jennifer Leung Schoenberger, DO
Department of Endocrinology, Rush University Medical Center, Chicago, IL, USA

Aisha Shaheen, M.D., MHA
Department of Surgery, Albert Einstein College of Medicine/Montefiore Medical Center, Bronx, NY, USA

Neha Shah Sheng, M.D.
Department of Vascular Surgery, Loma Linda University Medical Center, Loma Linda, CA, USA

David D. Shersher, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

George Singer, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Frederic Starr, M.D.
Department of Trauma, Stroger Cook County Hospital, Chicago, IL, USA

Ryan L. Steinberg, M.D.
Department of Urology, University of Iowa Hospitals & Clinics (UIHC), Iowa City, IA, USA

Gregory Stettler
Loyola University Stritch School of Medicine, Maywood, IL, USA

David Straus, M.D.
Department of Neurological Surgery, Rush University Medical Center, Chicago, IL, USA

Malini D. Sur, M.D.
Surgical Oncology, Department of Surgery, University of Chicago Medical Center, Chicago, IL, USA

Michael Tran, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Jacquelyn S. Turner, M.D.
Department of Surgery, Morehouse School of Medicine, Atlanta, GA, USA

Faaiza T. Vaince, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

Daniel Valentino, M.D.
Department of Surgery, Stroger Hospital of Cook County, Chicago, IL, USA

Benjamin Veenstra, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA

José M. Velasco, M.D.
Department of General Surgery, Rush University Medical Center, Chicago, IL, USA; Department of SurgeryNorthShore University HealthSystem, Skokie, IL, USA

Michael S. Vercillo, M.D.
Department of General Surgery, Cardiovascular and Thoracic Surgery, Rush University Medical Center, Chicago, IL, USA

Kaare J. Weber, M.D.
Department of Surgery, White Plains Hospital, White Plains, NY, USA

Elizabeth Weldon, M.D.
Obstetrics and Gynecology, Health System Clinician Northwestern University, Northwestern Memorial Hospital, Chicago, IL, USA

Part I
General Preoperative Considerations

1
Cardiac Clearance for Noncardiac Surgery

Ryan C. Knoper and Daniel Valentino

Purpose

The purpose of perioperative cardiac risk stratification is to determine cardiac risk level before proceeding with surgery. It requires that one order the appropriate testing and then intervene to best optimize the patient's status before surgery. This will decrease perioperative cardiac morbidity and mortality. A combination of surgical factors and patient factors must be considered (Fig. 1.1).

Surgical Factors

Surgical Timeline

Surgical procedures may be divided into emergent, urgent, and elective cases.

Emergency Surgery

For patients in critical condition who have a clear indication for emergent surgery, time will not permit cardiac risk stratification. For these patients invasive intraoperative monitoring should be considered, including Swan-Ganz catheters and intraoperative transesophageal echocardiography.

Urgent Surgery

Operations that need to be performed in a defined period of time may not allow for optimal medical management of comorbid conditions. In these situations there should be a collaborative discussion between the surgeon, anesthesiologist, and cardiology consultants regarding the patient's condition and the risks versus benefits of further cardiac evaluation and potential delays in surgery. In cases where surgery may be delayed a few days or weeks, patients should be tested and may undergo some procedures/therapy to improve their cardiac risk in the short term while not delaying their surgery for weeks to months as may be required for a revascularization procedure.

Elective Surgery

Lastly, for a scheduled elective procedure, a full cardiac risk evaluation should be performed to properly assess the patient's risk factors, and surgery should be delayed until the patient's comorbid conditions have been treated and/or optimized.

Risks of Surgery

Surgical procedures may be associated with different cardiac risk, independent of patient risk factors.

High Risk

Emergency surgery, peripheral vascular surgery (including aortic procedures), prolonged length of surgery, and large fluid shifts/blood loss.

Intermediate Risk

Intraperitoneal, intrathoracic, orthopedic, head and neck, and carotid endarterectomy.

Low Risk

Endoscopic, cataract, breast, and other superficial surgeries.

Patient Risk Factors

Multiple factors may predispose the patient to an increased cardiac risk level.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_1, © Springer New York 2015

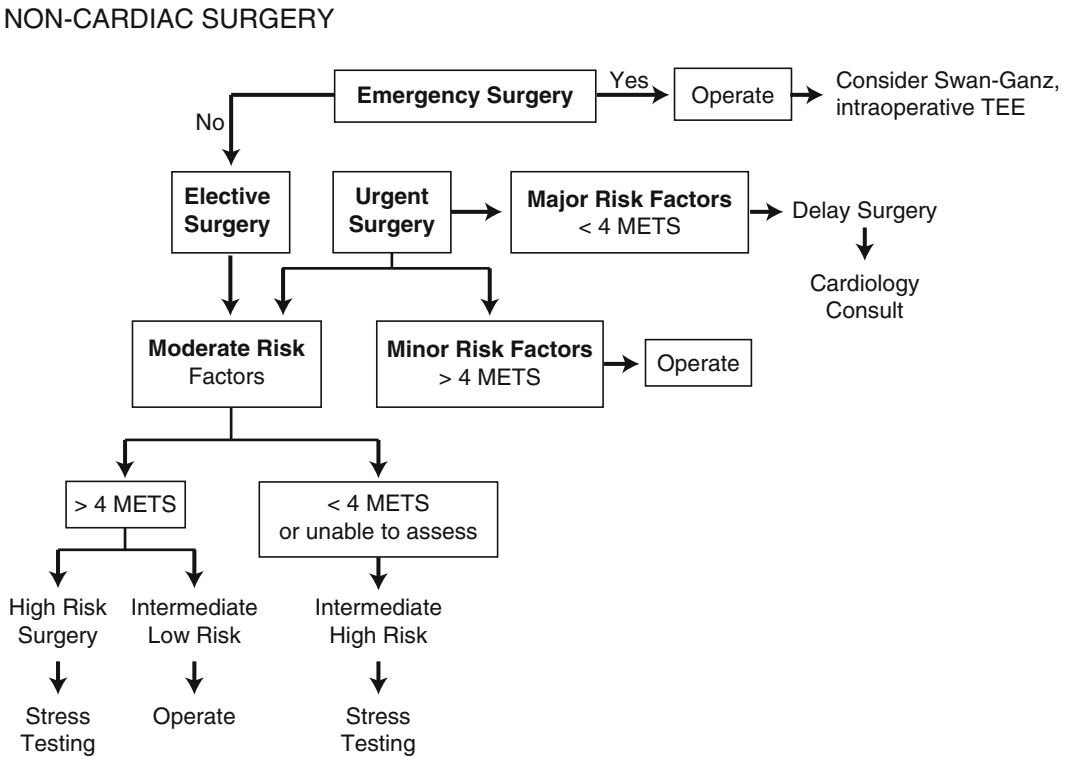

FIG. 1.1 Algorithm for assessing preoperative cardiac risk in patient undergoing noncardiac surgery. *MET* metabolic equivalent, *TEE* transesophageal echocardiography

Major Risk Factors

Acute or recent myocardial infarction (MI) with evidence of ischemia within 1 month of presentation, unstable angina, decompensated congestive heart failure (CHF), severe valvular disease, arrhythmias (including high-grade atrioventricular block, symptomatic ventricular arrhythmia, supraventricular arrhythmia with uncontrolled ventricular rate). *In these patients all elective surgery and urgent surgery should be delayed until the condition(s) are treated or optimized.*

Intermediate Risk Factors

Angina, previous MI (greater than 1 month prior to planned surgery), compensated CHF, diabetes mellitus, and renal insufficiency with creatinine ≥ 2 mg/dl.

Minor Risk Factors

Advanced age, non-sinus rhythm, hypertension, history of stroke, and reduced functional capacity defined in metabolic equivalents (Table 1.1).

Diagnostics

Patients undergoing low risk surgery with good functional capacity do not require further testing. For patients undergoing intermediate risk surgery, with good functional status, and have no more than two minor or one intermediate risk factors, further testing is usually not required. For patients undergoing intermediate- or high-risk surgery who have three or more minor and/or two or more intermediate-risk factors, in whom functional status is decreased, further testing is required. This includes assessment of ventricular function via echocardiography and/or stress testing. Pharmacologic stress testing is necessary for some patients. A positive stress test should be evaluated further.

Preoperative Optimization

Hypertension, especially stage III and above, should be managed. The treatment of valvular disease and heart failure remains the same as in the nonoperative setting. Severe or symptomatic cardiac disease may take precedence over urgent or elective surgery. In coronary artery disease, there is no evidence to support the use of percutaneous coronary intervention (PCI) to reduce perioperative cardiac morbidity in noncardiac surgery. The indications for PCI, coronary artery bypass

TABLE 1.1. Estimated energy requirements for various activities.

1 MET	Can you take care of yourself? Eat, dress, or use the toilet? Walk indoors around the house? Walk a block or two on level ground at 2 to 3 mph or 3.2 to 4.8 km per hour?
4 METS	Do light work around the house such as dusting or washing dishes? Climb a flight of stairs or walk up a hill? Walk on level ground at 4 mph or 6.4 km per hour? Run a short distance? Do heavy work around the house such as scrubbing floors or lifting or moving heavy furniture? Participate in moderate recreational activities such as golf, bowling, dancing, doubles tennis, or throwing a baseball or football?
Greater than 10 METs	Participate in strenuous sports such as swimming, singles tennis, football, basketball, or skiing?

MET indicates metabolic equivalent
Source: Adapted from the Duke Activity Status Index and AHA Exercise Standards

grafting, and cardiac catheterization remain identical to previously published ACC/AHA guidelines. Ideally noncardiac surgery should be delayed for 1 week after balloon angioplasty, 4–6 weeks after bare metal stenting, and 12 months after drug eluting stenting (DES). The cessation of antiplatelet therapy for any surgical procedure in patients who have a recent history of PCI should be done in consultation with a cardiologist to manage the risk of in-stent thrombosis and myocardial infarction.

Day of Operation

Patients should take their medications as prescribed for their cardiac risk factors with a sip of water. Exceptions include anticoagulation, antiplatelet, and diuretic medications. Beta-blockers should be taken the morning of surgery and continued postoperatively. Vitamin K antagonists should be stopped 5 days before surgery with appropriate bridging with low molecular weight or unfractionated heparin (UFH). UFH may be stopped 6 h and as close as 2 h before surgery, while low molecular weight heparin should not be given the night before surgery. Platelet inhibitors such as aspirin and clopidogrel should be stopped 7–10 days prior and may be restarted 24 h postoperatively. For those patients on antiplatelet therapy, platelet transfusion can be given in the perioperative period for emergent/urgent surgery. Pacemakers and implantable cardiac defibrillators (ICDs) should be turned off before surgery, via interrogation of the device by trained personnel.

2
Preoperative Evaluation of Bleeding

Lisa N. Boggio

Introduction

Bleeding is a major complication of surgical procedures. Bleeding leads to poor surgical outcomes, prolonged hospital stays, and increased postoperative complications (fluid overload, infections, etc.). Careful screening of patients preoperatively may identify patients who are at a higher risk of bleeding and may prevent further complications through a preoperative treatment plan.

Bleeding is caused by many factors. First the vessel wall needs to react to trauma. Nitric oxide and serotonin are released by the endothelium causing vasoconstriction. Platelets then plug the breach in the vessel wall, and clotting factors adhere to form a clot. When the endothelium is healed, the clot is removed through fibrinolysis, and the blood vessel dilates back to normal caliber. Abnormalities in any part of this process may lead to intraoperative bleeding.

Bleeding may be localized or diffuse. Diffuse bleeding from the operative site as well as from venipuncture sites is indicative of a systemic process such as a congenital or acquired coagulopathy. Most operative bleeding complications are due to a failure of local control in the operative field. Hypothermia, acidosis, and shock should be identified and corrected as soon as possible. A thorough preoperative evaluation can prevent many bleeding complications during surgery.

Preoperative Evaluation

A careful personal and family history should be obtained. Special attention should be given to any bleeding with previous surgical procedures. Obtain information about previous dental procedures as many patients do not consider this surgery and will not provide this information unless specifically asked. A family history of any bleeding disorder should be obtained as well as the patient's personal history of bruising, sensitivity to aspirin (ASA) or nonsteroidal anti-inflammatory drugs (NSAIDs), menorrhagia, or bleeding postpartum (Fig. 2.1). A history of any of these issues in the patient or any of their first-degree relatives should initiate a thorough bleeding disorder evaluation. Abnormalities of platelet number or function, prothrombin time (PT), partial thromboplastin time (PTT), and renal and liver function should be fully investigated. A personal history of liver and renal disease will increase a patient's bleeding risk. Patients with liver disease have a decrease in the levels of clotting factors and may have a decrease in platelet number. Renal failure increases bleeding risk through inhibition of platelet function.

A full list of all medications (Table 2.1), alcohol, and supplements that the patient is taking should be obtained. Alcohol not only causes a reduction in the coagulation factors though liver injury but also directly causes low platelet counts and leads to bleeding. Many herbs and compounds also cause bleeding (Table 2.2).

Antiplatelet agents need to be held for 5–7 days until their effect is reversed. The package insert for each medication should be consulted for specific recommendations of length of time to hold the medication preoperatively and when it is safe to resume postoperatively. Most low molecular weight heparins (LMWH) should be held for 24 h preoperatively and resumed 12–24 h post-op. Vitamin K antagonists (VKA) can be reversed emergently with fresh frozen plasma (FFP). For elective procedures, vitamin K at 2.5–5 mg orally can be given, but results will not be seen for 24 h after the vitamin K is given due to the half-life of the vitamin K-dependent clotting factors. Subcutaneous administration does not lessen the time for effect, and intravenous administration only decreases time to effect by a few hours with an additive risk of anaphylaxis. A goal INR of 1.5 or lower is considered safe for most surgical procedures; however this may increase the thrombotic risk, and some patients will need to be bridged with a LMWH. The newer anticoagulants do not have reversal agents, their

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_2, © Springer New York 2015

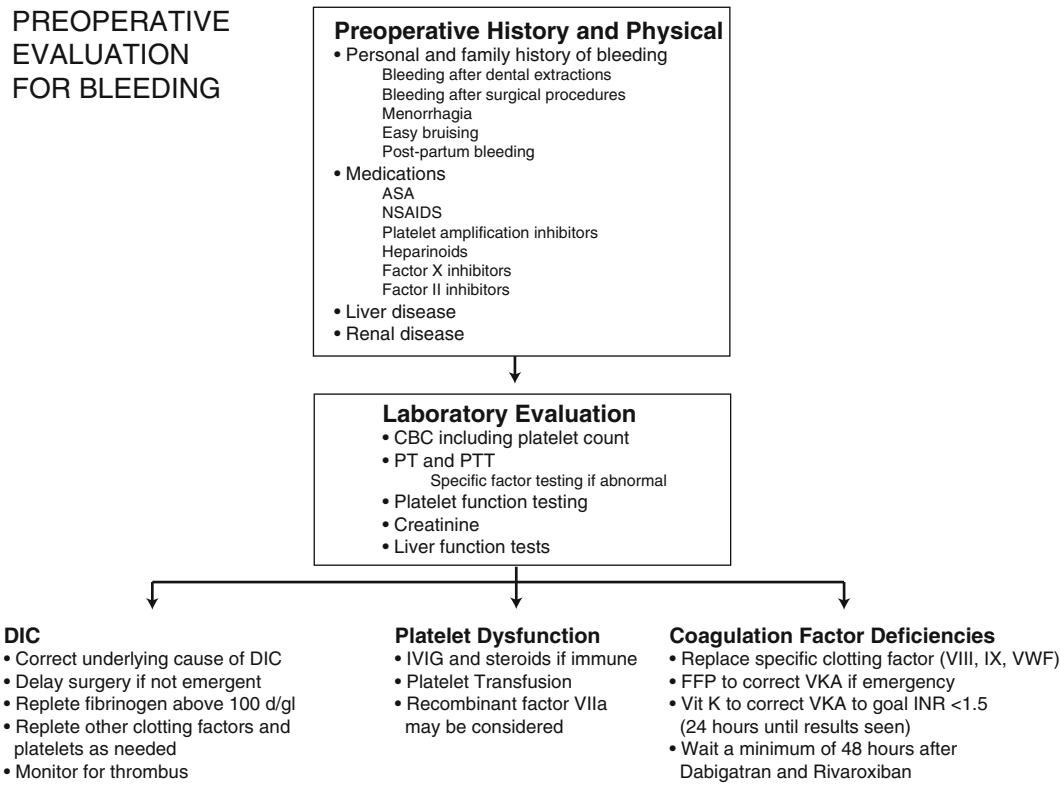

FIG. 2.1 Preoperative evaluation for bleeding. *ASA* aspirin, *NSAIDS* nonsteroidal anti-inflammatory drugs, *CBC* complete blood count, *PT* prothrombin time, *PTT* partial thromboplastin time, *DIC* disseminated intravascular coagulation, *IVIG* intravenous immunoglobulin, *FFP* fresh frozen plasma, *VKA* vitamin K antagonists, *INR* international normalized ratio, *VWF* von Willebrand factor

TABLE 2.1. Medications that cause bleeding.

Acetylsalicylic acid	Aspirin
NSAIDs	Ibuprofen
	Indomethacin
	Ketoprofen
	Ketorolac
	Naproxen
Platelet amplification inhibitors	Clopidogrel
	Meloxicam
	Ticlopidine
	Cangrelor
	Elinogrel
	Prasugrel
	Ticagrelor
Antidepressants	Citalopram
	Fluoxetine
	Fluvoxamine
	Paroxetine
	Sertraline
Vitamin K antagonists	Acenocoumarol
	Dicoumarol
	Warfarin
Heparins	Heparin
	Enoxaparin
	Dalteparin
	Tinzaparin
Factor X inhibitors	Fondaparinux (indirect)
Indirect	Rivaroxaban
Direct	Apixaban
Factor II inhibitors	Dabigatran

TABLE 2.2. Herbs and compounds that cause bleeding.

Ajoene	Birch bark
Black cohosh	Cayenne
Chinese black tree fungus	Cumin
Evening primrose oil	Feverfew
Garlic	Ginger
Ginkgo biloba	Ginseng
Grape-seed extract	Milk thistle
Omega-3 fatty acids	Onion extract
St. John's wort	Turmeric
Vitamins C and E	Willow tree bark

levels cannot be readily measured, and they have a high risk of inducing bleeding particularly in patients with renal impairment. They should be held a minimum of 48 h prior to procedures.

Disseminated Intravascular Coagulation (DIC)

Disseminated intravascular coagulation (DIC) is both a bleeding and thrombotic disorder. The clotting system becomes activated and forms thrombi or clots, and the clotting factors and platelets are trapped or "consumed" in the thrombi (consumptive coagulopathy). This causes a drop in the platelet number and activities of the clotting factors leading to bleeding. The identification of the underlying cause for DIC is essential for treatment as the DIC will wnot be resolved until the underlying cause is corrected. Common causes for DIC are infection, malignancy, liver disease, blood transfusion reaction, burns, head trauma, surgery, and anesthesia. DIC manifests as oozing from multiple sites, such as sites of previous venipuncture, purpura, and oozing from the operative site. Laboratory evaluation reveals a low platelet count, increased PT and PTT, an increased D-dimer, and low clotting factor activities – notably a low fibrinogen. Treatment is correction of the underlying cause and supportive care. Replace fibrinogen through infusions of cryoprecipitate, other clotting factors with fresh frozen plasma, giving platelet transfusions, and fluids/pressure support as needed to support the patient. Laboratory tests should be monitored frequently and supportive measures continued as needed.

Platelet Dysfunction

A decrease in platelet number (thrombocytopenia) is the most common acquired bleeding disorder. Thrombocytopenia may be immune (immune thrombocytopenic purpura, lupus), drug-induced immune reaction or bone marrow suppression (chemotherapy, antibiotics), dilutional after a large volume of fluids or blood, and bone marrow suppression (drugs, infection, malignancy) or due to sequestration in the spleen (liver disease, viral infections). Correction of the underlying cause preoperatively to optimize the platelet count is recommended. Steroids and intravenous immune globulin can acutely increase the platelet count in immune thrombocytopenia. Transfusions of platelets should be given after every 4–6 units of red blood cells infused during a massive transfusion protocol. The platelet count should be maintained at above 50,000/mcl in the perioperative period. Acquired platelet dysfunction due to antiplatelet agents can be treated with holding the medications for a week preoperatively, or, in the emergent setting, transfusion of platelets despite a normal platelet count may prevent excessive bleeding. Patients with platelet dysfunction due to renal disease may respond to desmopressin (DDAVP) given at a dose of 0.3 mcg/kg over 20 min (cap dose at 21 mcg). DDAVP may lead to hyponatremia so the sodium should be followed and free water limited after use.

Most platelet function defects have a normal number of platelets. Glanzmann thrombasthenia (glycoprotein IIb/IIIa defect causing a lack of platelet aggregation), Bernard-Soulier (GP1b/IX/V defect causing a lack of platelet adhesion), and storage pool disease (lack of procoagulant storage in platelets) are the most common platelet function defects. Treatment for bleeding is to transfuse platelets. Recombinant factor VIIa (rFVIIa) at a dose of 20 mcg/kg has been reported to have some effect in controlling bleeding in congenital platelet function defects.

Coagulation Factor Defects

Coagulation factor defects can be acquired due to decreased production from poor diet or liver disease (correction by vitamin K and/or FFP transfusion) or can be acquired from autoantibody inhibition—most commonly this is an autoantibody to factor VIII, which is treated with steroids and infusions of bypassing agents rFVIIa or prothrombin complex concentrates (PCC) to control bleeding. This is a rare disorder and consultation with an experienced hematologist is recommended.

Von Willebrand disease (VWD) is the most common bleeding disorder occurring in 1:100 persons. It is autosomal dominant and most patients have mild disease (type 1). Minor surgical procedures can be treated with DDAVP, but major surgical procedures require the use of a von Willebrand factor (VWF-containing product; Humate-P, Alphanate, Wilate). Types 2 and 3 VWD require the use of a VWF-containing product. For mucosal surgeries (dental procedures, tonsillectomy, uterine procedures), the addition of an antifibrinolytic (epsilon aminocaproic acid, cyklokapron) can aid in the control of bleeding. Antifibrinolytics should be avoided in bladder procedures due to the risk of obstruction.

Factors VIII (FVIII) and IX (FIX) deficiencies (hemophilias A and B) are X-linked recessive disorders, which occur in 1:5,000 and 1:15,000 males respectively. Males with mild FVIII deficiency can be treated with DDAVP for minor procedures, but major procedures require a FVIII concentrate. For major surgery the goal is to increase the factor activity to 100 % correction and to maintain a trough above 50 % for at least 2 weeks postoperatively. Inhibitors against FVIII or FIX (alloantibodies) occur in 30 % of those with FVIII deficiency and 3 % of FIX deficient patients. These patients do not respond to factor concentrates and must receive a bypassing agent such as rFVIIa or PCC. As these patients are complex, care at a hospital with a hemophilia treatment center is recommended.

Most hospitals do not carry factor concentrate, and patients are encouraged to bring their factor with them to

the hospital or the emergency department to ensure they get optimal care. FFP and cryoprecipitate are not optimal therapy in bleeding disorders as factor levels cannot be predictably raised, they are not virally inactivated, there is a large volume to be infused, and there are specific products for the treatment of bleeding in the hemophilias and VWD.

Conclusion

For any patient with a bleeding disorder, a thorough treatment plan should be obtained from his/her hematologist preoperatively. For any patient with a coagulopathy, communication between the surgeon and the hematologist preoperatively can help avoid complications.

3
Managing Patient's Medications (Diabetes, Beta Blockers, NSAIDs, Anticoagulants)

Shobha L. Rao

Introduction

Most patients undergoing surgery take medications on a daily basis. As our population ages, the number of patients on more than one chronic medication is increasing. Most of these medications are being taken for chronic medical populations, including hypertension, coronary artery disease, heart failure, and diabetes. Oftentimes, surgeons and internists need to decide which medications should be continued in the perioperative period and which medications should be discontinued. Within the literature, there is little outcome data about the vast majority of medications patients take in the perioperative period. The decision to continue or discontinue a medication is also often determined by the conflicting concerns of surgeons, anesthesiologists, and internists. Therefore, there are varied management strategies due to lack of medical evidence. This chapter will focus on some of the most common medications. The recommendations are based on expert opinion and meta-analyses.

Principles of Medication Management

- First and foremost it is imperative that a complete medication history that includes prescription, over-the-counter, and herbal supplements is obtained in the preoperative period. In addition, a complete social history that includes substance use, such as alcohol, tobacco, and illicit drugs, and the route of use should be performed. This should be updated immediately prior to surgery. All clinicians involved in the patient's care should carefully review this list.
- Medications that can cause morbidity or mortality if stopped abruptly or are not tapered should be continued or tapered if there is adequate time prior to surgery.
- Due to changes in splanchnic flow and fluid shifts, gastrointestinal absorption of medications may be impaired. If absorption will be altered due to decrease in gut motility or if the patient is to be kept NPO, alternate routes of administration should be considered (i.e., IV, transdermal, or transmucosal). If questions arise, it is important that the pharmacy be consulted.
- If medications are not necessary for a short period of time and may increase complications with anesthesia or surgery, they can be held during the perioperative period.

Nonsteroidal Anti-inflammatory Drugs (NSAIDs)

NSAIDs, in addition to providing pain control, are also antiplatelet agents, due to the reversible inhibition of COX-1, an isoform of cyclooxygenase. This leads to the decreased production of thromboxane A2, which is released by platelets, leading to platelet aggregation. This, in turn, can increase the perioperative risk of bleeding. NSAIDs are also implicated in renal failure, especially in the perioperative state, where intravascular volume status can be variable due to fluid loss during surgery and fluid shifts after surgery. Oftentimes, patients are also on ACE inhibitors, angiotensin receptor blockers, diuretics, and certain antibiotics, which can increase the risk of acute renal failure with concomitant use of NSAIDs. NSAIDs have also been implicated in increased adverse cardiovascular effects in patients who are at increased risk for cardiac complications.

Recommendations

Although NSAIDs can provide good pain control, it is recommended that NSAIDs be *discontinued* in the perioperative period due to increased risk for bleeding, acute renal failure, and adverse cardiac outcomes. Most NSAIDs should be discontinued at least 3 days prior to surgery to minimize the risk of perioperative bleeding. Ibuprofen can be used until 24 h prior to surgery. IV ketorolac is often used for pain control in the postoperative period for pain control, but this

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_3, © Springer New York 2015

should be used with caution. Patients should be well hydrated and renal function should be normal. Additionally, patients should not be on other nephrotoxic medications.

Beta Blockers

Beta blockers usually confer many benefits when taken in the perioperative period. Ischemia is decreased because beta blockers reduce myocardial oxygen demand secondary to increased release of catecholamines. Patients with stable angina who are dependent on their beta blocker are at increased risk for ischemia if discontinued abruptly. Beta blockers also aid in the prevention and/or control of arrhythmias that may occur. Significant morbidity and mortality is associated with the acute withdrawal of a beta blocker in the perioperative period as a patient can develop hypertension, tachycardia, and myocardial ischemia.

However, there are also risks associated with the use of beta blockers in the perioperative period, particularly bradycardia, hypotension, and potentially increased risk of stroke.

Recommendations

Because of the many benefits that outweigh the risks and the potential adverse effects from withdrawal, it is recommended that beta blockers should be continued in the perioperative period if it is a patient's chronic daily medication. The patient should take the beta blocker up to the time of surgery, including the day of surgery, and throughout the remainder of the hospitalization. The patient's blood pressure and heart rate should be monitored closely and titrated to prevent significant hypotension or bradycardia. There are recommendations that suggest reducing a patient's chronic beta blocker dose by half, starting the morning of surgery and for the first few days postoperatively. Also, if a patient's systolic blood pressure is less than 115 to 120 mmHg and the risk of sepsis is low, some experts recommend holding the beta blocker to prevent intraoperative and postoperative hypotension. This needs to be balanced with the risk of developing perioperative hypertension that can be associated with increased risk of stroke. Therefore, patients will need to be evaluated individually.

Beta blockers should be continued in patients with compensated heart failure, stable angina, atrial fibrillation, or a history of acute myocardial infarction. If patient has compensated heart failure but is not on a beta blocker and if surgery can be delayed for a few weeks, it is beneficial for this patient to be initiated on a beta blocker at least 4 weeks prior to surgery. However, if surgery cannot be delayed or is urgent, beta blockers should be initiated at some later time after surgery.

There is much discussion regarding whether the perioperative initiation of beta blockers can reduce cardiac complications, i.e., acute myocardial infarctions. Although there is benefit, there may also potentially be an increase of stroke as found in the POISE trial. Therefore, perioperative initiation of beta blockers is *not* recommended in patients with a low cardiovascular risk who will be undergoing low-risk noncardiac surgery. Initiation of a beta blocker is recommended in patients with coronary artery disease or ischemia noted on stress testing who will be undergoing vascular surgery, or in patients with more than one cardiac risk factor or CAD, who will be undergoing vascular surgery or intermediate risk surgery.

Beta blockers should not be started in patients with a baseline heart rate less than 60 beats/min, systolic blood pressure less than 90 mmHg, or when there is not enough time to safely titrate the dose to optimize medical management.

Which Route or Formulation to Use?

If a patient cannot take oral medications, IV metoprolol or labetalol can be used. Esmolol can be used if a patient is in the intensive care unit. Beta 1 cardioselective beta blockers are usually preferred due to the decreased risk for adverse pulmonary events or effects on the peripheral vascular system.

Anticoagulants

The risk of thromboembolism in patients on chronic anticoagulants needs to be weighed against the risk of bleeding when deciding if and when anticoagulation needs to be discontinued and if bridging is needed. The specific anticoagulation agent is also a factor. Many patients are on warfarin, which is a vitamin K antagonist, or low molecular heparin injections, such as enoxaparin or dalteparin. There are also new anticoagulants that are increasingly being used. Dabigatran is a direct thrombin inhibitor and rivaroxaban and apixaban are factor Xa inhibitors. These novel agents differ from warfarin as they have shorter half-lives and the lack of antidote or specific reversal strategy.

Estimate the Risk of Thrombosis and Bleeding

Risk of Thrombosis

- High Risk for Thrombosis – mitral valve prosthesis, caged-ball or tilting disk aortic valve prosthesis, recent stroke or TIA in the last 6 months, CHADS2 score of 5 or 6 in patients with atrial fibrillation, recent stroke or TIA in the last 3 months due to atrial fibrillation, rheumatic valvular heart disease, recent DVT/PE in the last 3 months, severe thrombophilia, e.g., protein C or S deficiency or antithrombin III deficiency, antiphospholipid antibody syndrome

- Moderate Risk for Thrombosis – bileaflet aortic valve prosthesis and one or more of the following risk factors: Atrial fibrillation, prior stroke or TIA, hypertension, diabetes, congestive heart failure, age >75; CHADS2 score of 3 or 4; DVT/PE in the last 3–12 months, recurrent VTE, active cancer
- Low Risk for Thrombosis – bileaflet aortic valve prosthesis without atrial fibrillation and no other risk factors for stroke, CHADS2 score of 0 to 2, VTE greater than 12 months in the past and no other risk factors for VTE

Procedural Bleeding Risks

- High-Risk Procedures – heart valve replacement, coronary artery bypass, AAA repair, neurosurgical/urologic/head and neck/abdominal/breast cancer surgery, bilateral knee replacement, laminectomy, TURP, kidney biopsy, polypectomy/variceal treatment/biliary sphincterectomy/pneumatic dilatation, PEG placement, endoscopically guided FNA, multiple tooth extractions, vascular and general surgery, and any major operation that lasts greater than 45 min
- Low Risk-Procedures – cholecystectomy, abdominal hysterectomy, GI endoscopy +/− biopsy, biliary/pancreatic stent without sphincterotomy, simple dental extractions, pacemaker and ICD insertion, carpal tunnel repair, knee/hip replacement and shoulder/foot/hand surgery and arthroscopy, dilatation and curettage, skin cancer excision, abdominal hernia repair, hemorrhoidal surgery, axillary node dissection, cataract and noncataract eye surgery, noncoronary angiography

Need for Bridging Anticoagulation

See Fig. 3.1.

Anticoagulation Agents

1. Warfarin

Warfarin is a vitamin K antagonist. It usually takes 2–3 days for the INR to fall below 2 and 4–6 days for the INR to normalize after stopping warfarin. The elimination half-life of warfarin is 36–42 h. It will also take about 4–6 days for the INR to become therapeutic after warfarin is restarted without heparin bridging. In patients where substantial blood loss is expected or has occurred, this may be the safest way to restart anticoagulation.

2. Dabigatran

Dabigatran is an oral direct thrombin inhibitor that is currently being used for stroke prevention in patients with atrial fibrillation. It does not require INR monitoring; however, it is expensive and insurance coverage needs to be assured prior to prescribing this drug. Its anticoagulant activity peaks 2–3 h after it is taken and has an elimination half-life of 12–15 h in patients with normal renal function and about 28 h in patients with severe renal dysfunction. Therefore, renal function needs to be monitored prior to restarting this drug after surgery as well as determining when the drug should be stopped prior to surgery.

- If the CrCl is greater than 50 %, stop dabigatran 1 to 2 days before the procedure.
- If the CrCl is less than 50 %, stop 3–5 days prior to the procedure.
- If that patient is undergoing major surgery, spinal puncture, or placement of a spinal or epidural port, it should likely be held for longer than the above durations.
- It is also recommended that an aPTT be documented at normal prior to surgery to ensure that dabigatran has been eliminated from the circulation.

Restarting following the procedure:

- Dabigatran can be resumed postoperatively once hemostasis has been reached.
- Given the rapid onset of action, surgeons need to be careful when restarting dabigatran, especially in patients with high bleeding risks. It may be prudent to wait 2–3 days after high-risk procedures prior to restarting dabigatran. If needed, a lower dose can be administered for the initial 2–3 days postoperatively or LMWH can be used.

Need for bridging anticoagulation:

- Because of the rapid onset and clearance of the dabigatran, bridging is often required.
- However, if the patient had been started on unfractionated heparin or LMWH after surgery and the patient needs to be transitioned to dabigatran, the dose should be given less than 2 h prior to the next dose of LMWH or at the time the heparin IV has been stopped.

3. Rivoroxaban and Apixaban

These two agents are oral direct factor Xa inhibitors. They also have a rapid onset of action with the peak of activity after 2–3 h. The elimination half-life is 9–13 h. These agents differ from dabigatran as they are less dependent on renal clearance. The above approach for dabigatran should be used for rivaroxaban and apixaban as well.

4. Low Molecular Weight Heparin (LMWH), i.e., Enoxaparin or Dalteparin

The half-life of subcutaneous LMWH is about 3–5 h. This drug cannot be used in patients with a CrCl less than 30 %. Also, usually patients heavier than 100 kg will need an internal medicine, hematology, or pharmacy consult as the distribution of the drug becomes less predictable.

- If a patient is on LMWH at home for anticoagulation, the patient's last dose should be 24 h prior to the planned surgery. It is recommended that if a patient is on a twice-daily

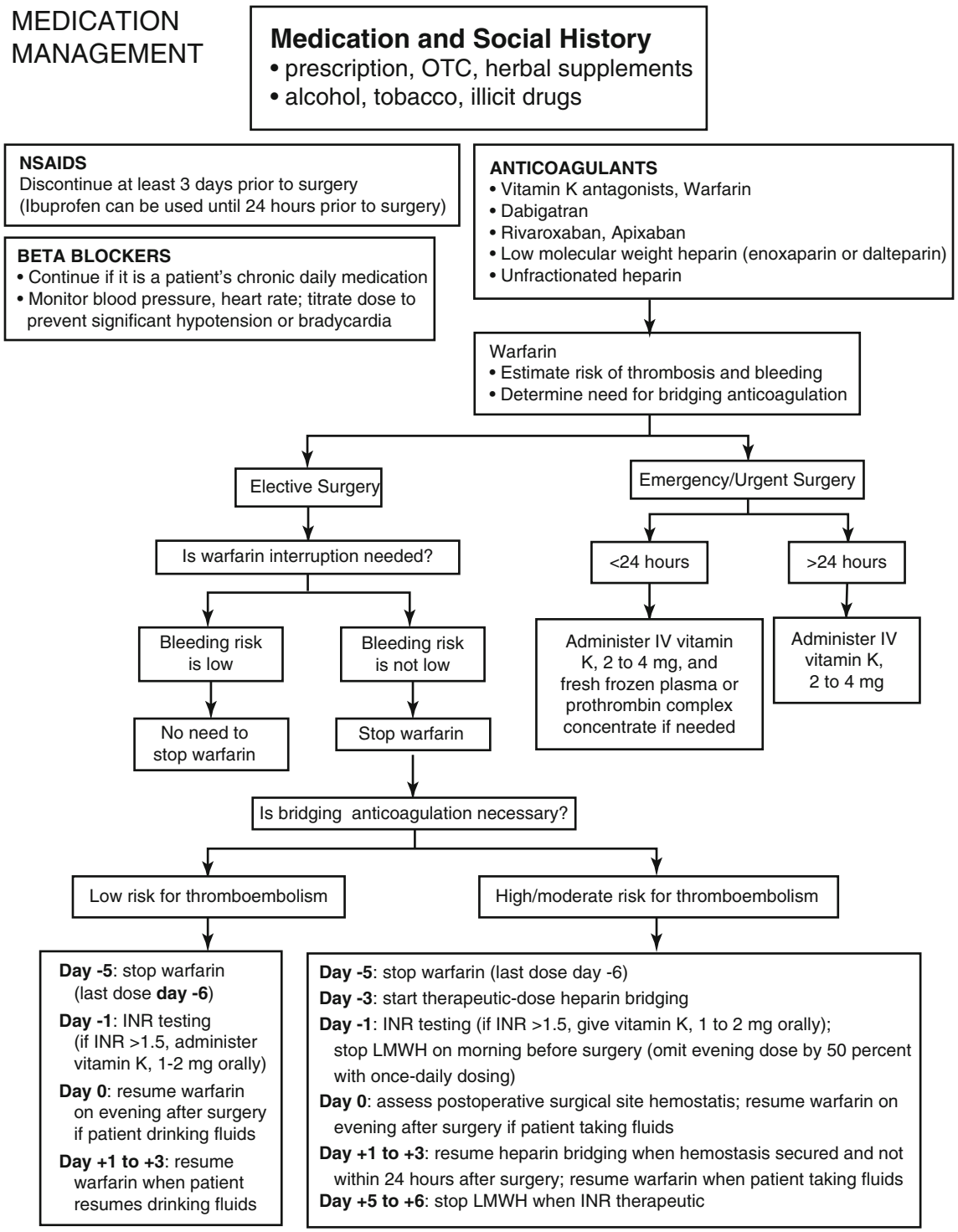

FIG. 3.1 Overview of perioperative management of patient's medications. *OTC* over the counter, *IV* intravenous, *INR* international normalized ratio, *LMWH* low molecular weight heparin (Adapted from Douketis JD. Perioperative management of patients who are receiving warfarin therapy: an evidence-based and practical approach. Blood 2011; 117:5044–5049)

dosing, the evening dose be held. If a patient is on a once-a-day dosing, the patient should take half of the normal dose the day before the procedure as some studies have shown residual anticoagulation effects after 24 h of stopping the dose.

- If there is concern that postoperative homeostasis has not occurred, LMWH should not be administered.
- LMWH can also be given at an intermediate dose, which is higher than the prophylactic dose for DVT prevention. For enoxaparin, the intermediate dose is 40 mg twice

daily, whereas the prophylactic dose to prevent DVT is 40 mg once daily.

5. Unfractionated Heparin

The half-life of IV unfractionated heparin is about 45 min. Usually, the drug is weight based and there is usually a published institutional algorithm to help dose heparin. In the perioperative setting, it is often not necessary to bolus the drug.

- Most studies suggest that the heparin drip be discontinued 4–5 h before the surgery or procedure.
- aPTTs need to be monitored closely and the drug will need to be adjusted based on the aPTT. UFH is usually considered therapeutic when the aPTT is 1.5 to 2 times the control aPTT.
- If a patient is considered high risk for bleeding postoperatively but requires bridging, UFH may be the safest way as the IV can be stopped immediately. There also exists an antidote that is not available for LMWH.

Diabetes Management

There is no consensus as to the best practice of glucose control, but there are several strategies that are accepted. However, none of these strategies optimally reduce morbidity, mortality, or length of hospitalization. Most experts agree on the following strategies.

The goals of diabetes management in the perioperative setting are as follows:

- Avoid hypoglycemia
- Prevent ketoacidosis
- Maintain fluid and electrolyte homeostasis
- Avoid profound hyperglycemia

Glucose readings should be between 140 and 200 mg/dL during the intraoperative and postoperative stages.

Preoperative Evaluation

- All patients should have a thorough preoperative history and physical performed. Emphasis should be placed on evaluating patients for microvascular and macrovascular complications of diabetes that could cause perioperative complications. Patients with diabetes are at increased risk for coronary heart disease, hypertension, chronic kidney disease, obesity, cerebrovascular disease, peripheral vascular disease, and neuropathy. All of these conditions pose risks with anesthesia and the postoperative period. Patients should also be asked about the frequency of hypoglycemic episodes.
- Every patient should get an ECG, a serum creatinine, blood glucose, and a hemoglobin A1c within the last 4–6 weeks.
- Ideally, all patients with diabetes should be scheduled to have their surgery prior to 9 AM to minimize the disruption of their medication regimen due to being NPO.

Perioperative Management

It is important to identify the patient as having Type 2 diabetes treated with diet alone, Type 2 diabetes treated with oral hypoglycemic medications/noninsulin injectables, or Type 1 or insulin-treated Type 2 diabetes. This will help determine the management of the patient's diabetes. The perioperative period is stress filled. Surgery and general anesthesia cause a neuroendocrine stress response that often results in hyperglycemia. The type of anesthesia, the extent of surgery, and other factors such as sepsis, glucocorticoid use, and hyperalimentation also influence glycemic balance, and this can often be unpredictable. Blood sugar levels are also influenced by vasopressor agents, hypotension, or critically illness. In these patients, venous or arterial blood testing should be used instead of finger-stick glucose levels.

Type 2 Diabetes Treated with Diet alone

- In general, these patients do not require aggressive therapy perioperatively.
- They may require intermittent doses of short-acting or rapid-acting insulin if their blood sugars rise over the optimal target.
- Blood sugars should be checked preoperatively and then again, soon after the completion of surgery. If the surgery is longer than 2 h or if it is complicated, which usually is associated with elevated blood sugar levels, blood sugars should be checked in the operating room every 1–2 h.

Type 2 Diabetes Treated with Oral Hypoglycemic Agents/Noninsulin Injectables

- Patients should hold their oral medications and noninsulin injectables on the morning of surgery as they are associated with certain adverse effects.
  - Sulfonylureas increase the risk of hypoglycemia, especially in patients with impaired renal function.
  - Metformin should not be restarted in patients with renal insufficiency, significant liver dysfunction, or congestive heart failure. Also, patients who received IV contrast for a CT scan are at risk for developing lactic acidosis in the presence of metformin.
  - Thiazolidinediones should not be used if there is risk for fluid overload as these can worsen edema and precipitate CHF.
  - Many of the noninsulin injectables can affect GI motility, which is often affected in the postoperative state.

- Patients with a hemoglobin A1c less than 7.0 are considered to have good glycemic control and therefore will likely not need insulin for short surgical procedures.
- If hyperglycemia develops, rapid- or short-acting insulin can be administered every 6 h.
- If a surgery will be longer than 2 h, again blood sugars should be monitored intraoperatively.
- Postoperatively, renal and hepatic function should be monitored closely prior to restarting oral hypoglycemics. Although these agents, except for metformin, can be started postoperatively, most experts would recommend not starting the oral or noninsulin injectables until the patient is taking adequate oral intake.

Type 1/Insulin-Dependent Type 2 Diabetes

- For minors, early morning procedures where only breakfast will be missed, patients may postpone their short- or rapid-acting insulin until after surgery and when they are able to eat.
- Patients who take long-acting insulin once daily (e.g., glargine or detemir) or use a continuous insulin pump should continue these meds as these are considered their basal insulin. If there is concern for hypoglycemia based on history or if a more conservative approach is preferred, the basal insulin dose can be reduced by 10 to 20%.
- If the patient takes intermediate-acting insulin, such as NPH, the dose should be reduced to one-half or two-thirds of the total insulin and given as intermediate- or long-acting insulin. The short-acting insulin should be omitted.
- If patients are going to have later surgeries, dextrose IV solution should be started at 75 to 125 cc/h to avoid ketosis or hypoglycemia.
- Blood sugars should be checked every hour and more frequently if blood sugars are less than 100.
- Long and complex surgeries will likely require an insulin infusion as studies have shown variability in blood sugars with subcutaneous insulin.
- Postoperatively, full dose subcutaneous insulin should not be initiated until patient is eating.
- If the patient is on an insulin infusion, it should be continued in patients who remain NPO after surgery. Once the patient is able to adequately take p.o., his/her first dose of subcutaneous insulin should be given prior to the discontinuation of the IV insulin infusion.
- If the patient was on subcutaneous insulin prior to surgery, he/she should be continued on a dextrose infusion until taking p.o. adequately.

Hypoglycemia

Hypoglycemia needs to be avoided as there are many adverse events associated with hypoglycemia. It is imperative in the intraoperative and postoperative period that patients are monitored closely as they will not be able to convey symptoms of hypoglycemia. Blood sugars need to be monitored aggressively in the operating room and postoperatively as well. Tremors, palpitations, anxiety, sweating, paresthesias, and hunger can all be manifestations of hypoglycemia. These can occur at glucose levels less than 70. As the hypoglycemia worsens, patients will develop cognitive and neurological impairment, including obtundation, seizures, and coma. If patients have poorly controlled blood sugars, these can occur at higher levels than anticipated.

Treatment

If the patient is awake and is not an aspiration risk, hypoglycemia can be treated with oral glucose in the form of tablet or gel or with juice. If patients are unable to take anything by mouth or are not awake, they should be given 25 g of 50 % dextrose IV. It is likely that an IV dextrose infusion may need to be initiated if the hypoglycemia does not resolve.

4
The Carotid Bruit

Erin Farlow

Background

Stroke is the third leading cause of death in the United States, and approximately 795,000 occur annually. Disease at the carotid bifurcation is responsible for 20–30 % of strokes.

National Stroke Association guidelines from the mid-1990s recommended basic blood pressure and pulse exams and auscultation for carotid bruit as screening tools for stroke prevention. Relying on the presence of a bruit, however, can be misleading. Only 25 % of asymptomatic patients with carotid bruits will have stenosis of 75 % or greater when they undergo additional evaluation. Also, patients who have high-grade stenosis or occlusion will not present with a bruit, making auscultation a screening tool with poor sensitivity and specificity (Fig. 4.1).

Those at increased risk for stroke include older patients with a history of other vascular disease, high cholesterol, diabetes, hypertension, and a history of smoking. While general ultrasound screening does not have proven cost effectiveness, ongoing studies are evaluating the potential benefit of focused ultrasound screening in high-risk populations.

Evaluation

For patients with suspected disease at the carotid bifurcation, evaluation should begin with a *bilateral* carotid duplex ultrasound. Approximately 50 % of patients with carotid stenosis will have a contralateral lesion that may factor into the final treatment plan. For most patients, ultrasound exam by a well-trained technician is adequate to assess the length and degree of stenosis for operative planning.

CT angiogram (CTA) may play a role when there is concern for concomitant vertebrobasilar insufficiency or when stenting is being considered over open operative repair. CTA evaluates the tortuosity of the arch and takeoff of the carotids to see if stenting is feasible.

While diagnostic carotid angiograms were formerly the gold standard for evaluating carotid disease, they are now infrequently performed given that one-third of perioperative strokes associated with carotid endarterectomy (CEA) were thought to be secondary to the angiogram.

Medical Management

Medical treatment of carotid atherosclerosis involves managing and minimizing the risk factors – smoking cessation, hyperglycemic control, and management of hyperlipidemia and hypertension. In addition, antiplatelet agents have been extensively studied. *Studies have shown that daily aspirin leads to a 22 % decrease in the incidence of stroke*. There does not seem to be an increased benefit with higher doses, so 81 mg or 325 mg daily provides adequate protection. The addition of clopidogrel has not decreased the rate of stroke; however, it has increased bleeding complications.

Carotid Endarterectomy

The American Heart Association currently recommends that *patients with asymptomatic disease and 60–99 % stenosis undergo CEA provided the individual surgeon stroke/mortality rate is less than 3 %*. These recommendations are supported by the Asymptomatic Carotid Atherosclerosis Study (ACAS), which showed that patients with greater than 60 % stenosis when treated with CEA versus aspirin alone had a reduced risk of death or stroke (5 % versus 11 %) over a 2.7-year follow-up.

When patients are symptomatic with previous TIA or stroke within 6 months and have 70–99 % stenosis, CEA is recommended provided the surgeon has less than a 6 % rate of stroke/mortality perioperatively. This is supported by the North American Symptomatic Carotid Endarterectomy Trial (NASCET) in 1991, which showed that symptomatic patients with stenosis greater than 70 % treated with CEA over medical treatment alone had a 9 % versus a 26 % incidence of stroke over a 2-year period.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_4, © Springer New York 2015

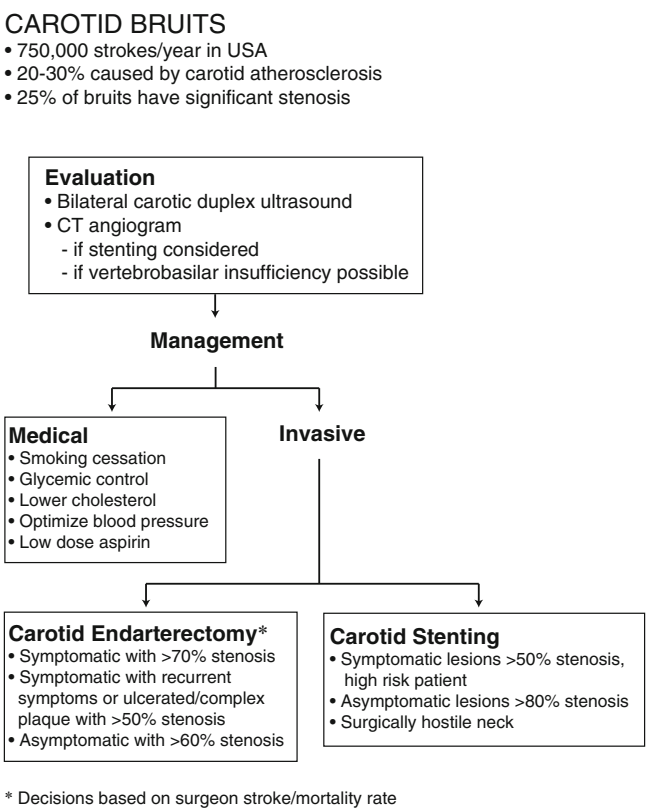

FIG. 4.1 Algorithm for treatment of carotid bruit. *CT* computed tomography, *CEA* carotid endarterectomy

Additionally, symptomatic patients with 50–69 % stenosis may be considered for CEA if their lesions are extensive or irregular in appearance or if they have recurrent symptoms despite appropriate medical management.

After surgery is recommended, additional intra-op decision-making is required. Once the dissection has been performed, the surgeon must evaluate the need for intraoperative shunting. While performed regularly in some institutions, risks associated with it include plaque embolization or dissection of the proximal internal carotid with introduction of the shunt. Therefore, most institutions use shunts selectively for patients who appear to have inadequate perfusion otherwise (approximately 20 % of patients). Indications for intraoperative shunting include a back bleeding pressure of less than 40 mmHg, EEG changes with clamping of the internal carotid, changes in somatosensory evoked potential, or decreased transcranial Doppler blood flow in the ipsilateral middle cerebral artery.

The endarterectomy itself can also be performed by two techniques. The standard technique uses a longitudinal incision along the carotid onto the internal carotid. The plaque is then gently peeled from the media, and the arteriotomy is closed either primarily or with a small vein or bovine pericardium patch if the artery was felt to be at risk for stenosis with primary closure. Eversion is the second technique whereby the internal carotid is transected at its origin and the atheroma is intussuscepted from the artery. If the internal carotid was found to be tortuous, it can be shortened with this technique and then closed circumferentially. Complications of CEA include hypoglossal, accessory, or vagus nerve injury; infection; hyperperfusion syndrome; and pseudoaneurysm.

Carotid Artery Stenting (CAS)

Ongoing studies are evaluating the appropriate role for carotid artery stenting (CAS). Currently, it should be considered in patients less than 80 years with favorable anatomy, high cardiac risk, those with recurrent lesions, or those with a history of cervical radiation or cervical dissection for malignancy. The FDA has approved CAS for those at high risk when symptomatic with greater than 50 % stenosis or those that are asymptomatic with greater than 80 % stenosis. CAS must be considered carefully in patients with marginal renal function given the IV dye required.

Post-op Management

Patients undergoing CAS should receive clopidogrel for 1 month following procedure and aspirin for life. They should have a duplex exam at 2–4 weeks, 6 months, and yearly after the procedure. Patients with CEA should be monitored at 6 months and yearly following.

5
DVT Prophylaxis

Neha Sheng

Introduction

Deep venous thrombosis (DVT) prophylaxis is an important clinical issue since its complications are grave. The incidence of DVT is 10–40 % in medical and general surgical patients. The complications of venous thromboembolism (VTE), including pulmonary embolism (PE) and post-thrombotic syndrome (PTS), cause significant morbidity and mortality. *Pulmonary embolism is the most common cause of preventable death in patients admitted to the hospital*, causing 150,000–200,000 deaths per year in the United States. The incidence of fatal PE in the absence of prophylaxis is estimated to be 0.1–0.8 % after elective general surgery, and as high as 7 % in emergency orthopedic surgery. Post-thrombotic syndrome is manifested by a constellation of symptoms caused by venous hypertension in patients who have sustained DVT, and may include edema, pain, varicose veins, and venous ulceration.

The American College of Chest Physicians (ACCP) guidelines for DVT, 9th edition, clearly state that hospitals should employ a formal DVT prevention protocol. Compliance rates for DVT prophylaxis are below 60 %, but can be improved with mandatory clinical protocols. The ideal protocol is safe, effective, easy to execute and cost effective. The individual patient's risk of VTE should be assessed and then a plan chosen based on this risk. Prophylaxis can be undertaken with chemical, mechanical, or a combination of methods.

Risk Assessment

The pathophysiology for DVT is based upon *Virchow's triad*: stasis, hypercoagulability, and endothelial injury. Overall, surgical patients have a twofold increased risk of VTE versus medical patients. Both patient-related and procedure-related risk factors exist. Table 5.1 includes a comprehensive list of patient-related risk factors. Risk factors related to the specific operation performed include patient positioning (e.g., reverse Trendelenburg position impairs venous return), the use of a tourniquet and the length of the operation.

Several acceptable regimens of mechanical and chemical prophylaxis for DVT are described in the American College of Chest Physicians official recommendations. The choice of the appropriate regimen is dependent upon the assessment of risk specific to that patient and procedure.

Patients are categorized as follows: low, moderate, high, and very high risk of VTE (Table 5.2). Low risk patients include patients with no additional risk factors for thromboembolism, undergoing minor surgical procedures, and in need of no more than 30 min of general anesthesia. These patients may only require early and frequent ambulation for the prevention of VTE. Younger patients with risk factors, or undergoing major surgery, and older patients without additional risk factors comprise the moderate risk category. High and highest risk groups include patients with multiple risk factors and older patients.

Lastly, the patient's risk of bleeding must be assessed, especially in the surgical patient. If the surgeon deems the risk of bleeding to be too high for chemical VTE prophylaxis, only mechanical methods should be employed. The ACCP guidelines note that most minor foot and ankle surgery, vascular surgery, and laparoscopy do not require routine VTE prophylaxis.

The incidence of VTE is progressively increased with each of these categories of risk. Proximal DVT has greater morbidity than distal DVT. Incidence of distal DVT ranges from 2 % in the low risk group, to 10–20 % in the moderate, 20–40 % in high, and as high as 40–80 % in the highest risk group. The incidence of proximal DVT is lower in the low risk group – it is 0.4 %, increasing to 2–4 % in the moderate, 4–8 % in the high, and 10–20 % in the highest risk groups. Fatal PE is the most feared complication of DVT. It is very rare in the low risk group. The incidence of fatal PE in the moderate risk group is 0.1–0.4 %, increasing to 0.4–1 % in the high and 0.2–0.5 % in the highest risk groups. See Table 5.2 for a summary of the likelihood of VTE in each risk group.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_5, © Springer New York 2015

Table 5.1. Risk factors for DVT.

Hospitalization
Recent surgery or trauma
Malignant neoplasm
Pregnancy and postpartum period
Age
Previous central venous catheter or pacemaker
Previous superficial venous thrombosis
Neurologic disease with extremity paresis
Varicose veins
Congestive heart failure
Blood group A
Inflammatory bowel disease
Oral contraceptive pill use or hormone replacement therapy
Iliac vein compression
Paroxysmal nocturnal hemoglobinuria
Selective estrogen receptor modulators
Inflammatory bowel disease
Nephritic syndrome
Myeloproliferative disorders
Acute medical illness
Obesity
Smoking
Air travel > 6 h
Inherited thrombophilia
Prothrombin G20210 mutation
Factor V Leiden mutation
Protein C deficiency
Protein S deficiency
Dysfibrogenemia
Elevated clotting factors
Elevated homocysteine
Lupus anticoagulant and anticardiolipin antibodies.
Heparin-induced thrombocytopenia
Fibrinolysis
Behcet's syndrome
Lupus
Homocysteinuria
Popliteal vein entrapment
Venous aneurysms
Inferior vena cava anomalies
Risk factors for recurrent DVT
Obesity
Older age
Malignant neoplasm
Extremity paresis
Superficial venous thrombosis

Regimens for DVT Prophylaxis

Several acceptable regimens for DVT prophylaxis exist. They include either mechanical prophylaxis, chemical prophylaxis, or both. Mechanical prophylactic devices include intermittent pneumatic compression devices (IPC) and graduated compression stockings (GCS). Chemical prophylaxis may include unfractionated heparin (UFH), low molecular weight heparin (LMWH), fondaparinux, or vitamin K agonists (VKA). Heparin bonds with antithrombin and inactivates multiple factors in the coagulation cascade. Unfractionated heparin is injected subcutaneously in increments of 5,000 units every 8 or 12 h. There is no definitive data that addresses whether dosing every 8 or 12 h is more safe or effective; hence, either regimen is acceptable. It is generally accepted that higher dosing is required for obese and high risk patients. Low molecular weight heparins, such as enoxaparin or dalteparin, activate antithrombin. Acceptable dosing for enoxaparin includes 30 mg subcutaneously injected twice daily, or 40 mg daily. The dosing of dalteparin is 2,500 or 5,000 international units (IU) injected subcutaneously daily. *The main advantage of low molecular weight heparin is a lower incidence of heparin-induced thrombocytopenia*. Fondaparinux is an inhibitor of factor Xa, and can be used in VTE prophylaxis; the usual dosing is 2.5 mg injected subcutaneously every day. *Neither LMWH nor fondaparinux should be administered for patients with renal failure and creatinine clearance less than 30 ml/min*, without consultation with a nephrologist. Warfarin is an oral vitamin K agonist that is used only in special cases for VTE prophylaxis. Dose adjustment of warfarin to keep the INR between 2 and 3 is required. Aspirin, clopidogrel, ticlopidine, idraparinux, rivaroxiban, and dabigatran are anticoagulant and antiplatelet agents, which have no indications for prevention of DVT in surgical patients.

For low risk patients, early and frequent ambulation is adequate prophylaxis for DVT. For patients at moderate risk, LMWH, UFH, GCS, and IPC are all viable options, starting preoperatively and continuing until the patient begins to ambulate. High and highest risk patients should receive a combination of mechanical and chemical thromboprophylaxis. Please refer to the algorithm in Fig. 5.1 for selection of the most appropriate regimen for each risk group.

Special Considerations

Chemical VTE prophylaxis should be initiated 2 h prior to the operation. The recommended duration of prophylaxis is until discharge for most patients. However, some special circumstances exist for particularly high risk patients:

1. In the case of major surgery for cancer, prior VTE, or morbid obesity surgery thromboprophylaxis should be continued up to 28 days postoperatively.
2. For patients undergoing hip or knee arthroplasty, or an operation for hip fracture, prophylaxis should be administered for a minimum of 10 days and up to 35 days. These patients are at especially elevated risk for VTE. LMWH, starting 12 hours before surgery, is recommended in this population. IPC should be added to this regimen while the patient is in the hospital.
3. In the setting of trauma and spinal cord injury, LMWH has been shown to provide better prophylaxis than UFH. In patients with spinal cord injury, chemical VTE prophylaxis should be delayed until hemostasis is complete. Only mechanical methods of thromboprophylaxis should be used until hemostasis is evident. This will prevent progression of injury.
4. Fatal PE is the most common cause of postoperative death in the bariatric population, ranging between 0.2 and

TABLE 5.2. Risk assessment for VTE.

Risk level	Low	Moderate	High	Very high
	Under age 40 without additional risk factors	Age 40–60 without additional risk factors	Age >60 without additional risk factors	Age >60 undergoing major surgery or with additional risk factors
	Minor surgery/general anesthesia <30 min	Age <40 undergoing major surgery without additional risk factors, *or* minor surgery with additional risk factors	Age 40–60 undergoing major surgery without additional risk factors, *or* minor surgery with additional risk factors	Age 40–60 undergoing major surgery *and* with additional risk factors
		General anesthesia >30 min *and* no additional risk factors	Age <40 undergoing major surgery with additional risk factors	Age <40 with multiple additional risk factors, undergoing major surgery
			Includes most orthopedic surgery, colorectal surgery, major trauma, spinal cord injury, or cancer surgery	
Risk of proximal DVT	0.4 %	2–4 %	4–8 %	10–20 %
Risk of distal DVT	2 %	10–20 %	20–40 %	40–80 %
Risk of PE	0.2 %	1–2 %	2–4 %	4–10 %
Risk of fatal PE	<0.01 %	0.1–0.4 %	0.4–0.1 %	0.2–0.5 %

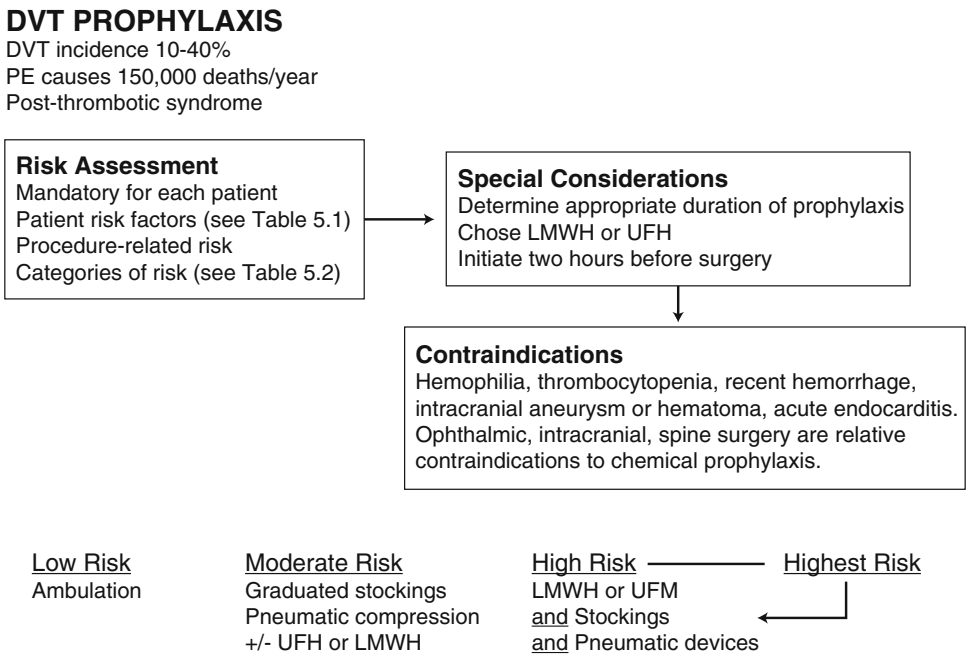

FIG. 5.1 Treatment algorithm for DVT. *PE* pulmonary embolism, *LMWH* low molecular weight heparin, *UFM* unfractionated heparin

0.3 %. Chemical thromboprophylaxis at higher doses is recommended in bariatric patients, as long as bleeding risk is acceptable.

Prophylactic use of inferior vena cava filter (IVCF) for prevention of VTE has no definite indications. However, some clinicians utilize this procedure in patients at very high risk, especially when chemoprophylaxis is contraindicated. Prophylactic IVCF has been described for surgical patients who are critically ill, have morbid obesity, hypercoagulable state, prolonged immobility, malignancy, spinal cord injury, multiple trauma (especially multiple orthopedic injuries), intracranial hemorrhage, and ocular injury. We await higher quality data from randomized prospective trials, and treatment should be individualized at this time.

Contraindications

Risk of bleeding should be evaluated by the surgeon, and decisions regarding chemoprophylaxis for VTE should be individualized for each patient. The contraindications to anticoagulant medications preclude chemoprophylaxis and include the following: hemophilia or other bleeding disorders, thrombocytopenia (especially with platelet count

less than 70,000 platelets/cubic millimeter), active or recent hemorrhage, intracranial aneurysm or hematoma, and acute bacterial endocarditis. Recent intracranial, ophthalmic, or spine surgery are some relative contraindications. It is safe for these patients to undergo mechanical thromboprophylaxis with IPC and/or GCS. IPC is absolutely contraindicated in patients with existing DVT, but may be used on the contralateral limb if no DVT exists in that limb. In the presence of severe peripheral arterial disease, IPC and GCS are relatively contraindicated due to the risk of skin necrosis from compression.

Conclusion

In conclusion, VTE prophylaxis is one of the most important patient safety measures. *Hospitals should employ mandatory protocols for screening for risk of VTE and implementation of appropriate regimens for prophylaxis.* All patients should undergo screening with a thorough history and physical examination. All patients should also have some form of DVT prophylaxis during their hospital stay, ranging from ambulation, to mechanical and chemical forms of prophylaxis.

References

1. Arcelus JI, Caprini JA. "Acute Deep Venous Thrombosis: Prevention and Medical Treatment." Rutherford's Vascular Surgery. Ed. Cronenwett JL, Johnston KW. Philadelphia:Saunders, 2010. 770–791.
2. Gould MK, Garcia DA, Wren SM, Karanicolas PJ, Arcelus JI, Heit JA, Samama CM. Antithrombotic Therapy and Prevention of Thrombosis, 9th ed: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines. Chest 2012;141(2) (suppl):e227s–277s.

Part II
Trauma

6
Initial Trauma Assessment and Resuscitation

Frederic Starr

Introduction

In the United States, accidental injury is the leading cause of death for people between the ages of 1 and 44. Traumatic injury of all causes is responsible for more than 170,000 deaths annually. More than 42 million people will seek medical care due to intentional or accidental trauma this year, at an annual national cost of greater than $406 billion dollars.

The injured patient must be assessed quickly and treatment of life-threatening injuries begun immediately. A systematic approach to this evaluation assures that the most critical injuries are identified early and that potentially lethal injuries are not missed. The American College of Surgeons Advanced Trauma Life Support (ATLS) course promotes the "ABCDE" primary survey sequence, and this is the most widely accepted approach to the initial evaluation of the trauma patient (Fig. 6.1). This strategy also provides a framework for reevaluation if the patient's condition deteriorates, redirecting the physician back to the start of the algorithm in search of a missed or worsening injury.

Once the primary survey is complete and the patient's vital signs are normalizing, the secondary survey is begun.

Preparations

Prior to the patient's arrival, the hospital trauma team should be mobilized, including notification of radiology, blood bank, respiratory therapy, and the operating room. Intravenous (IV) fluids should be warmed, and the trauma bay should be warmed if possible. The team should follow standard precautions (cap, gown, gloves, mask, shoe covers, face shield).

Airway

Evaluation of the patient's airway is the first priority. Spontaneous speech in an awake patient indicates a patent airway. Depressed level of consciousness may cause airway obstruction due to collapse of the soft pharyngeal tissue or blockage by the posterior movement of the tongue, a situation that can be improved with a jaw thrust or chin lift maneuver, or placement of an oral airway. Similarly, severe facial trauma may lead to obstruction, mandating immediate intervention. Indications for intubation include inability to protect the airway, profound shock, or a Glasgow Coma Scale (GCS) less than or equal to 8. Nasotracheal intubation requires a spontaneously breathing patient and is contraindicated in patients with severe facial trauma or suspected basilar skull fractures. The orotracheal route is generally the preferred method for airway control. A contingency plan should exist for the difficult airway and includes adjuncts such as bougies, Combitubes, and intubating laryngeal mask airways (LMAs). Video laryngoscopes are generally of limited use in the acute trauma setting as the equipment is often not immediately available, and the presence of blood or gastric contents in the oropharynx limits visibility. Inability to intubate mandates a surgical airway, with cricothyroidotomy the preferred procedure in adults. Needle cricothyroidotomy is favored in children younger than 12 years old due to their narrow cricoid ring, but it is important to note that this temporizing measure only provides oxygenation, not ventilation. A definitive airway will need to be secured either by more experienced personnel or surgically in the operating room. All patients with blunt head trauma or altered mental status should be presumed to have a cervical spine injury until proven otherwise. Inline cervical spine stabilization should be maintained at all times when securing a definitive airway.

Breathing

Once the airway is assessed and secured, all trauma patients should be started on supplemental oxygen. Breathing first should be evaluated by inspection, looking for external signs of injury, asymmetry of chest rise, paradoxical motion, and the use of accessory respiratory muscles. Tracheal deviation or the presence of distended neck veins should be noted.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_6, © Springer New York 2015

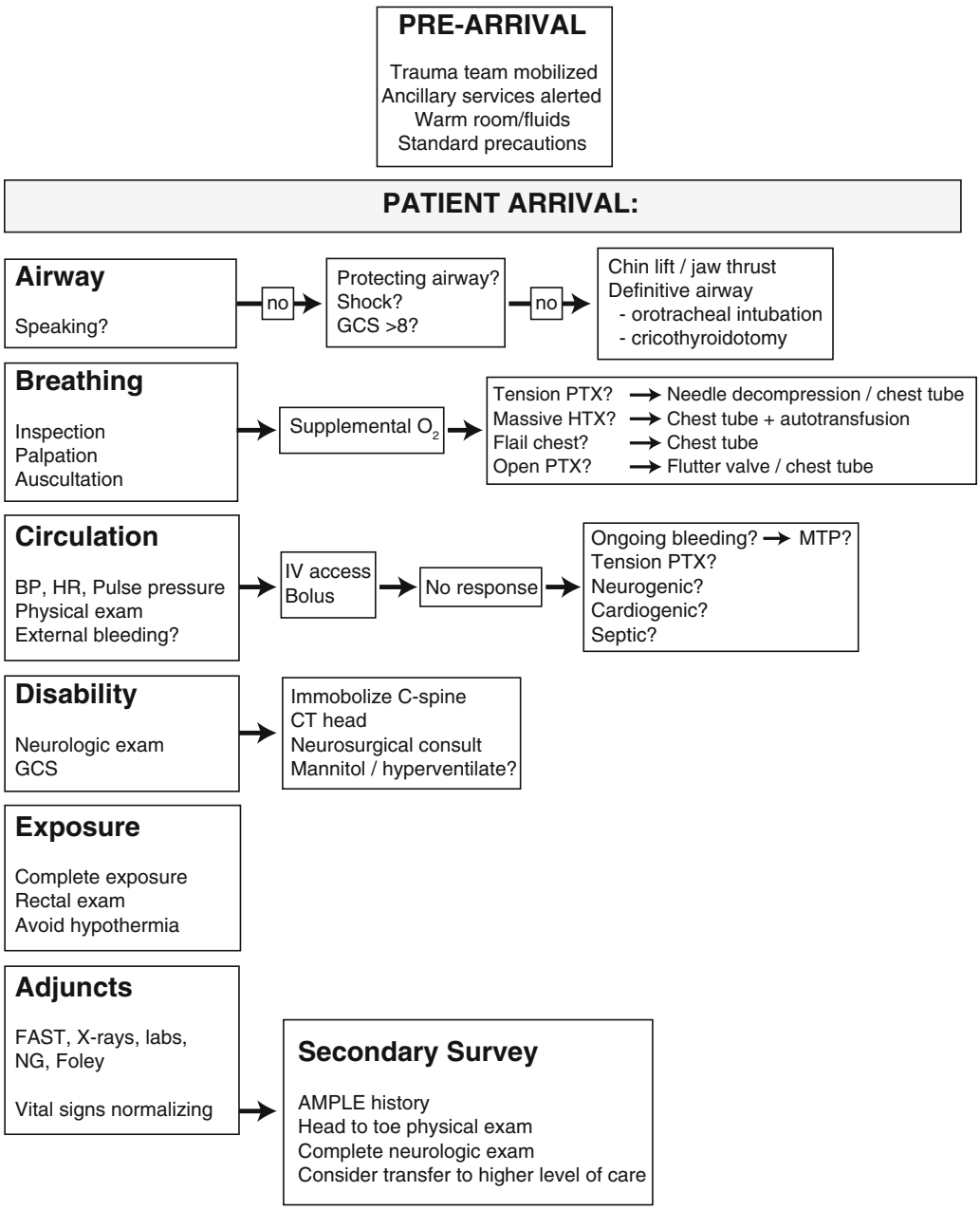

FIG. 6.1 The "ABCDE" primary survey sequence (airway, breathing, circulation, disability, exposure) for assessing and treating trauma, followed by a secondary survey when the patient's vital signs begin to normalize. *GCS* Glasgow Coma Scale, *PTX* pneumothorax, *HTX* hemothorax, *BP* blood pressure, *HR* heart rate, *IV* intravenous, *MTP* massive transfusion protocols, *CT* computed tomography, *FAST* focused assessment with sonography for trauma, *NG* nasogastric, *AMPLE* – A = allergy, M = medications, P = past medical/surgical history, L = last meal, E = events of injury

The chest should also be palpated to identify areas of tenderness or subcutaneous emphysema. Percussion may illicit hyperresonance or dullness, indicating pneumothorax or hemothorax, respectively, though in a noisy trauma bay this may be impractical. Finally, auscultation may demonstrate absent or diminished breath sounds from a hemo-/pneumothorax or malpositioned endotracheal tube. Life-threatening injuries that must be identified and addressed during the primary survey include tension pneumothorax, massive hemothorax, flail chest, and open pneumothorax. A tension pneumothorax should be suspected in a hypotensive patient with absent breath sounds, hyperresonance, distended neck veins, and deviated trachea. Treatment is immediate needle decompression (before the chest X-ray!)

with a large-gauge angiocatheter through the second intercostal space in the midclavicular line. Massive hemothorax is defined as greater than 1,500 mL of blood within the pleural space, and initial treatment requires prompt chest tube placement (36–40 Fr). Adding an autotransfusion canister to the chest tube drainage system should be considered. Flail chest occurs with segmental fractures in three or more adjacent ribs and raises concern for underlying pulmonary injury as well as resultant hypoventilation secondary to pain. Treatment is supportive, occasionally requiring mechanical ventilation. Lastly, an open pneumothorax ("sucking chest wound") occurs with chest wall defects that are greater than 2/3 the diameter of the trachea, resulting in air flowing predominantly through this new path of least resistance. A flutter valve should be created using an occlusive dressing taped on 3 sides, allowing air to flow out but not in with each breath. A chest tube must be inserted and ultimately the defect closed.

Circulation

Shock, defined as inadequate organ perfusion and tissue oxygenation, can be categorized as hemorrhagic, neurogenic, cardiogenic, or septic. Hypotension in a trauma patient is due to blood loss until proven otherwise, and the degree of hemorrhage can be estimated quickly by physical exam. Level of consciousness, skin color, pulse rate, and pulse pressure rapidly reflect volume status. Systolic blood pressure, however, usually does not fall until 30 % of the blood volume is lost (class 3 hemorrhagic shock) and therefore may deceive one into a false sense of security. The most important concept is *locate and stop the bleeding*. External bleeding must be identified and controlled, usually with direct pressure. Other possible locations of hemorrhage include the thorax, abdomen, retroperitoneum, and extremities. Intravenous access must be established, generally with two 16 gauge (or larger) antecubital IVs, and a 2 l bolus of lactated Ringer's solution is given. Triple lumen catheters should be avoided due to their high resistance to flow (long tube and small diameter). Instead, a 7.5 French Cordis is preferable, generally placed in the femoral vein. If the patient improves hemodynamically, the fluids are decreased to a maintenance rate. If there is no response to the initial bolus, one must consider transfusing blood (O negative or type-specific). Many trauma centers have developed massive transfusion protocols (MTP) that facilitate rapid delivery of large quantities of blood to the exsanguinating patient. Not only are packed red blood cells (PRBCs) provided but also fresh frozen plasma (FFP) and platelets. Recent literature reports improved survival if administering these blood products in a 1:1:1 ratio. In addition to bleeding, other potential causes of shock must be entertained. Tension pneumothorax has been discussed. Neurogenic shock is characterized by hypotension in the face of a normal heart rate (or even bradycardia due to unopposed vagal cardiac stimulation). It occurs with spinal cord injury above the mid-thoracic level and *not* brain injury, and treatment consists of fluids initially, followed by vasopressors if necessary. Pericardial tamponade is generally associated with penetrating injuries and can be recognized by hypotension, tachycardia, jugular venous distension (JVD), and muffled heart sounds. Echocardiography is the mainstay of diagnosis. Treatment is immediate operation, though pericardiocentesis can stabilize the patient until the operating room is available or transfer to an appropriate facility can be achieved. Cardiogenic shock can also be due to blunt cardiac injury, which is diagnosed by EKG and echocardiogram. Treatment is supportive. Septic shock is extremely rare in the acute trauma setting.

Disability

Brain or spinal cord injury can be detected by a brief neurologic exam. Before any paralytic agents are given for intubation, movement of all four extremities should be assessed and lateralizing signs noted. Abnormal pupillary exam, including size and reactivity, can indicate intracranial injury. The Glasgow Coma Scale is the most widely used assessment of level of consciousness and incorporates the best exam score in three categories: eye opening, verbal response, and motor response. Scores range from 3 (worst) to 15 (best). One advantage of using the GCS is its reproducibility and simplicity, allowing frequent reevaluations by different physicians. All patients with suspected head injury require emergent noncontrast head computed tomography (CT) for rapid diagnosis and differentiation of operative and nonoperative pathology. Severe head injury with elevated intracranial pressure and signs of herniation is treated initially with mannitol and MILD hyperventilation ($PaCO_2$ 30–35). More vigorous hyperventilation is discouraged and leads to cerebral vasoconstriction, limiting cerebral oxygen delivery. Cervical spine injury must be assumed with all blunt trauma, and cervical immobilization with a C-collar is employed until bony and ligamentous injury can be ruled out. Currently, the use of steroids for suspected or confirmed blunt spinal cord injury is discouraged by all major neurosurgical and orthopedic societies.

Exposure

The patient should be completely exposed (including rolling) and the entire body examined for signs of injury. A rectal exam should be performed. To avoid hypothermia, the patient should then be covered with warm blankets once the inspection is complete. Again, IV fluids and blood should be warmed.

Adjuncts

During the primary survey, blood pressure, EKG, and pulse oximeter monitors should be placed. Supplemental oxygen should be administered. If clinically indicated, a nasogastric tube and Foley catheter are inserted. Basic imaging can also be obtained at this point, including chest, pelvis, and lateral cervical spine radiographs. Focused assessment with sonography for trauma (FAST) is often included at the end of the primary survey and is particularly useful in multisystem blunt trauma patients with hypotension of unknown origin. FAST exam allows rapid detection of hemopericardium and hemoperitoneum while, in the trauma bay, sparing the unstable patient from a potentially disastrous trip to the CT scanner. Extended FAST is also being used to evaluate for pneumothorax and hemothorax.

Secondary Survey

Once the primary survey is completed and the patient's vital signs are normalizing, a thorough head-to-toe exam is performed. A complete neurologic exam is performed. A brief "AMPLE" history (A = allergy, M = medications, P = past medical/surgical history, L = last meal, E = events of injury) should be taken. Any change in the patient's condition mandates reassessment of the primary survey. If the patient's injuries require management not immediately available at the current hospital, transfer to a higher level of care must be considered and arrangements initiated. Diagnostic studies should not delay transfer to definitive care.

7
Head Trauma

David Straus

Initial Resuscitation

Initial assessment and management of the head trauma victim involve securing the patient's airway, breathing, and circulation. Moderate to severe traumatic brain injury (TBI) is associated with ~5 % incidence of concomitant cervical spine injuries, and maintenance of spinal immobilization is necessary, particularly in the obtunded patient. Nasogastric intubation in patients with severe craniofacial trauma or evidence of skull base fractures risks intracranial misplacement of the tube. One should promptly correct hypoxia ($SpO_2 < 90$ %), hypoventilation, and hypotension (SBP <90 mmHg) that occur at any point during the initial resuscitation to minimize risk of secondary brain injury. A focused medical history (including usage of anticoagulant/antiplatelet medicines) and a thorough understanding of the mechanism of injury and the energy involved in the accident are essential.

Neurological Evaluation

The baseline neurologic exam is determined only *after* the patient is completely resuscitated, as cardiopulmonary insufficiency will impact the neurologic exam. One should note if and when the patient has received sedating medications or paralytics (especially in recently intubated patients). The immediate post-resuscitation *Glasgow Coma Scale* (GCS) (Table 7.1) has significant prognostic implications, functions as an important baseline of neurologic function, and should be documented in all head trauma patients. Head trauma may be clinically stratified based on the GCS as follows: severe TBI (GCS 3–8), moderate TBI (GCS 9–12), or mild TBI (GCS 13–15). Patients with severe TBI ($GCS \leq 8$) may require intubation. A *complete* neurologic examination – assessing mental status, pupillary responses, cranial nerves, brainstem reflexes, segmental motor function, sensation, reflexes, and coordination should be obtained. Secondary survey assesses the presence of raccoon eyes, Battle sign, hemotympanum, otorrhea, rhinorrhea, periorbital ecchymosis/edema, palpable skull fractures/defects, and scalp lacerations. The complete absence of brainstem reflexes may preclude further intervention and should be noted.

Laboratory Evaluation

Testing is aimed at detecting conditions that may confound or exacerbate neurological deficits. Urine and serum toxicology screens, urine pregnancy screen, aPTT, INR, CBC, BMP, ABG, serum glucose, and crossmatch are indicated. Coagulopathies, hypercapnia, hypoglycemia, and other metabolic abnormalities should be aggressively corrected. One should maintain platelets >100,000, INR <1.3-1.5, Hgb >7.0–8.0, and serum sodium at the upper end of normal (~145 mmol/L). Platelet transfusion should be considered in patients with intracranial hemorrhage and a history of antiplatelet use regardless of platelet count.

Radiographic Evaluation

Close observation without imaging is acceptable for patients with GCS 15, without loss of consciousness, focal neurological deficit, or clinical concern for underlying brain injury. In all other head trauma patients, noncontrast computed tomography (CT) of the brain is indicated as the initial exam. Early imaging is crucial to determine the severity, anatomy, and pathology of the underlying traumatic injury and, in conjunction with the neurologic examination, forms the basis of therapeutic decision making. Important findings on CT include presence of skull fractures, mass lesions, subarachnoid or intraventricular hemorrhage, compression/absence of basal cisterns, and the amount of midline shift.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_7, © Springer New York 2015

Table 7.1. Glasgow Coma Scale.

Eye opening	E4	Open spontaneously
	E3	Open to voice
	E2	Open to pain
	E1	Closed
	E1c	Unable to evaluate (swelling/injury)
Verbal	V5	Oriented
	V4	Confused
	V3	Incoherent
	V2	Incomprehensible
	V1	Nonverbal
	V1t	Unable to evaluate (intubated/injury)
Motor	M6	Follows commands
	M5	Localizes
	M4	Withdraws to pain
	M3	Flexor posturing
	M2	Extensor posturing
	M1	No response to pain
Total score: E + V + M (range 3–15)		

Table 7.2. ABC/2 volume measurement.

ABC/2 method	
A	Largest diameter of clot (cm)
B	Largest diameter of clot perpendicular to A (cm)
C	Count 1 cm CT slices above and below where clot is present (75–100 % of maximum size = 1, 25–50 % = 0.5, 0–25 % = 0)
Estimated volume of clot (cc) = (A x B x C)/2	

Penetrating Head Trauma

Penetrating injuries are either stab-type injuries or gunshot wounds (GSW). Mortality from intracranial GSW is high: 48 % if a single lobe is involved, 72 % in multilobar injuries, 77 % if the midsagittal plane is crossed, 84 % if the midcoronal plane is crossed, and 96 % if both the midsagittal and coronal planes are crossed. In addition to CT, vascular imaging (either conventional cerebral angiogram or CT angiogram) is indicated in penetrating trauma. Surgical interventions for penetrating intracranial injuries are necessary in salvageable patients. Goals of surgical intervention include debridement of devitalized scalp, skull, dura, brain tissue, and accessible foreign bodies. Decompressive craniectomy may or may not be required depending on the severity of the underlying brain injury. Surgical reconstruction of the scalp, watertight dural closure, and repair/cranialization of injured air sinuses are critical to minimize the risk of cerebrospinal fluid (CSF) leak and infection. Prophylactic antiepileptics and broad-spectrum antibiotics are indicated for all penetrating cranial injuries.

Blunt Head Trauma

Management of blunt head trauma is based on the underlying pathology. Injuries are categorized as diffuse or focal injuries. Often multiple injuries are present in a single patient. Severe TBI has ~33 % incidence of intracranial hematoma, moderate TBI has ~8 % risk, and mild TBI has ~0.2 % incidence. *Diffuse injuries* consist of concussions, cerebral edema, traumatic subarachnoid hemorrhage, diffuse axonal injury, and intraventricular hemorrhage. Diffuse blunt head injuries are treated with supportive measures and intracranial pressure management (see below). Surgical interventions for diffuse injuries are limited to placement of intracranial pressure (ICP) monitors, brain oxygen monitors, ventriculostomy placement for ICP monitoring, and/or CSF diversion and decompressive craniectomy for management of refractory ICP. *Focal injuries* include epidural hematomas (EDH), acute subdural hematomas (SDH), intracerebral hemorrhage (ICH), contusions, and posterior fossa hematomas. Surgical indications for evacuation of focal injuries rely on neurologic exam, volume of hematoma measured on CT, and degree of midline shift (MLS) (Table 7.2). *Epidural hematomas* most frequently occur from lacerations of meningeal arteries but may also result from calvarial fractures or venous sinus lacerations. They appear as convex lesions that do not cross suture lines on CT.

Nonsurgical management requires serial CT and neurological exams: the risk of EDH expansion is significant, particularly with an arterial source of bleeding. *Acute subdural hematomas* are associated with severe brain injury and typically result from tearing of bridging cortical veins. They appear as a hyperdense concave lesions that transverse suture lines and may layer along the falx or tentorium. *Traumatic contusions* and *intracerebral hematomas* are complex lesions that frequently evolve ("blossom") after the initial injury. Surgical evacuation of intra-axial hematomas is more dangerous than evacuation of extra-axial lesions due to the risk of intraoperative and postoperative bleeding and the difficulty in differentiating hematoma from viable contused brain. Surgical indications and techniques vary.

Surgical Management of Focal Injuries

Emergent surgical evacuation of an EDH is indicated if any of the following findings are present: clot volume ≥30 cc, clot thickness ≥15 mm, MLS ≥5 mm, GCS <9, or focal

neurological deficit. Surgical thresholds are lowered for epidural hematomas in the middle cranial fossa due to the increased risk of transtentorial herniation and resultant brainstem compression. Emergent surgical evacuation of acute SDH is indicated if the clot thickness is ≥10 mm, MLS is ≥5 mm, post-resuscitation GCS declines by ≥2 points, and ICP is >20 mmHg or if the patient is anisocoric or has fixed and dilated pupils. Published guidelines for surgical intervention for intra-axial hematoma includes progressive neurologic deterioration referable to the intraparenchymal lesion, refractory intracranial hypertension, signs of mass effect on CT, ICH volume >50 cc, patients with GCS 6–8, *and* frontal or temporal contusions >20 cc *and* MLS >5 mm or cisternal compression. Surgical options for ICH include clot evacuation, subtemporal decompression, decompressive lobectomy (frontal/temporal), decompressive hemicraniectomy, and decompressive bifrontal craniectomy. Patients with posterior fossa hematomas are at significant risk of rapid and profound neurologic deterioration. Emergent surgical evacuation of posterior fossa hematomas is indicated in the presence of any neurological deficit or deterioration referable to the lesion, if there is evidence of mass effect (obstructive hydrocephalus, effacement/distortion of the fourth ventricle) or if there is cisternal compression.

Other Head Injuries

Closed (simple) *skull fractures* may be elevated with the intention of improved cosmesis, fracture healing and posttraumatic seizure reduction if the fracture is depressed >10 mm or greater than the thickness of the adjacent skull. Open skull fractures should be repaired to prevent infections and posttraumatic seizures. Operative indications include depressed fracture greater than the thickness of the surrounding skull, dural penetration, pneumocephalus, frontal sinus involvement, wound infections, gross contamination, or cosmetic deformity. *Air sinus fractures* may require cranialization of the involved sinus to prevent infection, particularly if both anterior and posterior tables are fractured or if there is an associated dural defect or CSF leak. *Skull base fractures* most commonly involve the petrous temporal bone. They may result in injury to the cranial nerves (particularly the facial and vestibulocochlear nerves), the petrous segment of the internal carotid artery injury or CSF leaks. *Traumatic CSF fistulae* predispose to meningitis; treatment includes prophylactic antibiotics during active otorrhea/rhinorrhea. Interventions to repair the fistula progress from flat bed rest, to lumbar drain placement, to endoscopic repair and/or craniotomy.

Extracranial *dissections of the internal carotid or vertebral arteries* typically result in ischemia from embolism and are usually treated with anticoagulation; decision making may be complicated by the presence of intracranial hemorrhage. Intracranial dissections typically result in subarachnoid hemorrhage and often require surgical or endovascular intervention. Head injuries can cause *traumatic intracranial aneurysms. Dural arteriovenous fistula* and *carotid-cavernous fistula* may also arise as a consequence of head trauma. *Cerebral venous sinus lacerations* may occur in skull fractures overlying a sinus and may require repair. *Venous sinus thrombosis* may also occur in the setting of head injury and may require anticoagulation. Posttraumatic hydrocephalus requires ventriculoperitoneal shunting.

Nonoperative Management

Supportive care of the head trauma patient revolves around preventing secondary brain injury. In any significant head trauma, ICU-level care and close physiologic and invasive monitoring are necessary. Local and/or diffuse brain swelling peaks between 3 and 5 days post-injury. *Intracranial pressure management and support of cerebral perfusion are central to the care of TBI*. Invasive ICP monitoring should be instituted in patients with severe TBI (GCS below 9). ICP and mean arterial pressure (MAP) should be managed to maintain cerebral perfusion pressure (CPP = MAP – ICP) between 50 and 70 mmHg. *ICP should be kept below 20 mmHg*. Early interventions include maintaining neutral neck position, elevating the head of bed (30°), and ensuring adequate analgesia and sedation and normocarbic ventilation ($PaCO_2 \sim 35$ mmHg). Second-tier medical interventions include mannitol and hypertonic saline (serum osmolality should not exceed 320 osm). Second-tier surgical intervention includes ventriculostomy drainage of CSF. Third-tier medical therapy may include mild to moderate hypothermia (<24 h), moderate hyperventilation (not to exceed $PaCO_2 < 25$ mmHg), or barbiturate coma (titrated to burst suppression on EEG). Third-tier surgical intervention is limited to decompressive craniectomies. Occasionally, brain oxygen monitoring will be instituted via jugular venous sampling (target $SjvO_2 > 50$ %) or brain tissue monitoring (target $PbrO_2 > 15$ mmHg).

Blood pressure is monitored continuously; MAP is supported by maintaining euvolemia (CVP 8–12) and with vasopressors as necessary to maintain CPP in 50–70 mmHg range. Episodes of hypotension (SBP <90 mmHg) and hypoxia ($SpO_2 < 90$ %, $SaO_2 < 60$ mmHg) are avoided.

Episodes of hypoglycemia are injurious as is persistent hyperglycemia. Prophylactic antiepileptics are administered for 7 days. Steroids for TBI are associated with worsened outcomes and are contraindicated. Standard infection prevention measures should be instituted, and fevers should be aggressively treated with antipyretics and cooling as necessary. Fluid and serum electrolyte status should be carefully monitored and maintained in normal ranges. TBI may result in cerebral salt wasting, SIADH, or diabetes insipidus. Full caloric support should be instituted by post-injury day 7 at the latest. H2 blockers are used for GI ulcer prophylaxis; mechanical DVT prophylaxis is necessary at all times.

Conclusion

Head trauma is a significant cause of mortality and morbidity. Management of the head trauma patient requires standard primary trauma survey, immediate post-resuscitation neurological evaluation and head CT, emergent surgical intervention when indicated, supportive care to minimize secondary brain injury, and rehabilitation following the acute injury (Fig. 7.1). Important prognostic variables include mechanism of injury, cardiopulmonary stability, age, post-resuscitation GCS, and radiographic findings. Recovery and long-term prognosis may not be certain until 1 year after the injury.

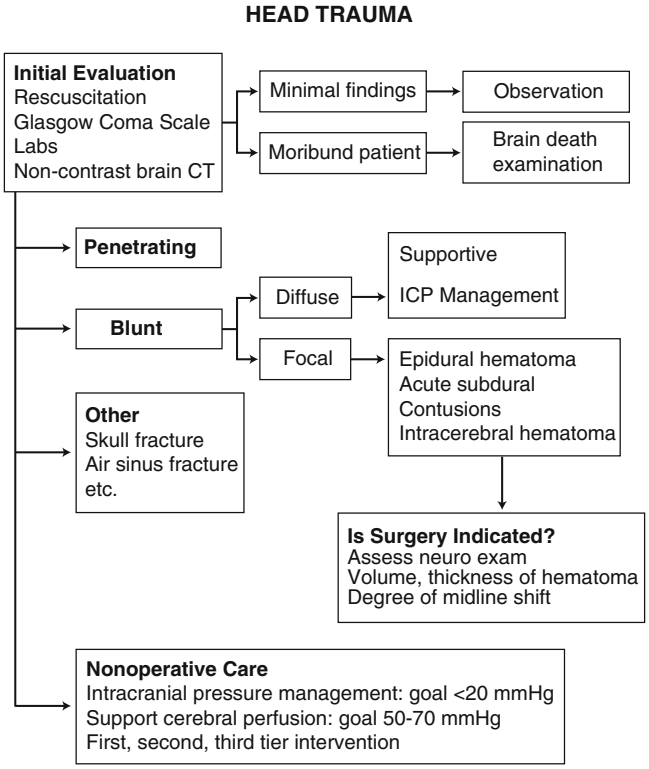

FIG. 7.1 Head trauma treatment algorithm

8
Management of the Difficult Airway

Phillip S. LoSavio

Introduction

A universally accepted definition for the "difficult airway" can be challenging to define based on the current literature. The American Society of Anesthesiologists (ASA) describes it as "the clinical situation in which a conventionally trained anesthesiologist experiences difficulty with face mask ventilation of the upper airway, difficulty with tracheal intubation, or both." A variety of protocols, mnemonics, and guidelines have been developed to aid a physician in managing these difficult and challenging cases. As with any basic or advanced life support, the establishment of an adequate airway and ventilation are the first and foremost steps in stabilizing a patient's vital sign parameters. Inadequate ventilation will ultimately lead to a rapid decline in oxygenation and subsequent arrest. Further efforts to resuscitate will prove fruitless without taking the time to stabilize this basic and core aspect of the patient's physiology.

Airway Management

In reality, a difficult airway is a complex situation with many interrelating factors. The patient, the experience of the physician, and the clinical setting all come into play when deciding on the best course of action. In the modern era, the anesthesiologist in most hospitals will take the lead role in managing airways. Fortunately, difficult laryngoscopy is reported in only 1.5–8.5 % of general anesthesia procedures with less than 1 % having a failed intubation. Advanced noninvasive airway techniques have been introduced over the past decades that continue to facilitate intubation and ventilation by the anesthesiologist. Ultimately, though, the surgeon serves as the last hope of rescuing a patient who cannot be ventilated by noninvasive means and does so by performing a cricothyrotomy or tracheostomy. It is important for the surgeon to have a firm understanding of the full scope of airway management, especially as it relates to trauma patients and those with head and neck surgical pathology.

The history is the first step in evaluating a patient. In the case of airway management, unless this is an elective same-day surgery, a quick clinical assessment of the patient's status takes precedent and should be done first and foremost. The physician must quickly assess the patient's vitals and ability to spontaneously ventilate on his or her own. If this is not the case, resuscitation will need to begin, and a rapid response team called to assist in the situation. Simultaneously one needs to assess his or her own skills and decide if further help will be required. In addition, one needs to quickly assess what technical limitations are present in the current care setting. Are you on the floor in the middle of the night moonlighting as the only house officer or in the operating room at 9 AM on a weekday? Assuming the patient is stable and this is an elective case where immediate action is not necessary, you can begin by taking a thorough history of the patient. A past history of intubation problems in the medical record should alert the physician to potential difficulty. Factors such as obstructive sleep apnea, history of neck radiation, or prior neck or cervical spine surgery are examples of factors that should alert the physician of a potential problem with airway management.

There is insufficient evidence to point toward any specific anatomic exam features that predict the feasibility of noninvasive airway access. There are a multitude of identified risk factors and rating systems that exist in the literature. One of the most known and regarded is the Mallampati classification scale that grades tongue size and visualization of the soft palate (see Fig. 8.1). Other factors include occlusion (over- or underbite), trismus (ability to open mouth the width of three fingers), thyro-mental distance, and neck length/range of motion/size. Other exam features would also include pathologic features such as neck or tongue swelling, tumor, voice changes, or stridor.

Advanced thoughtful planning if possible will inevitably lead to better outcomes in most difficult clinical scenarios. The same holds true for airway management. A basic plan set out between the surgeon and anesthesiologist a few minutes in advance of the case will allow for a more controlled

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_8, © Springer New York 2015

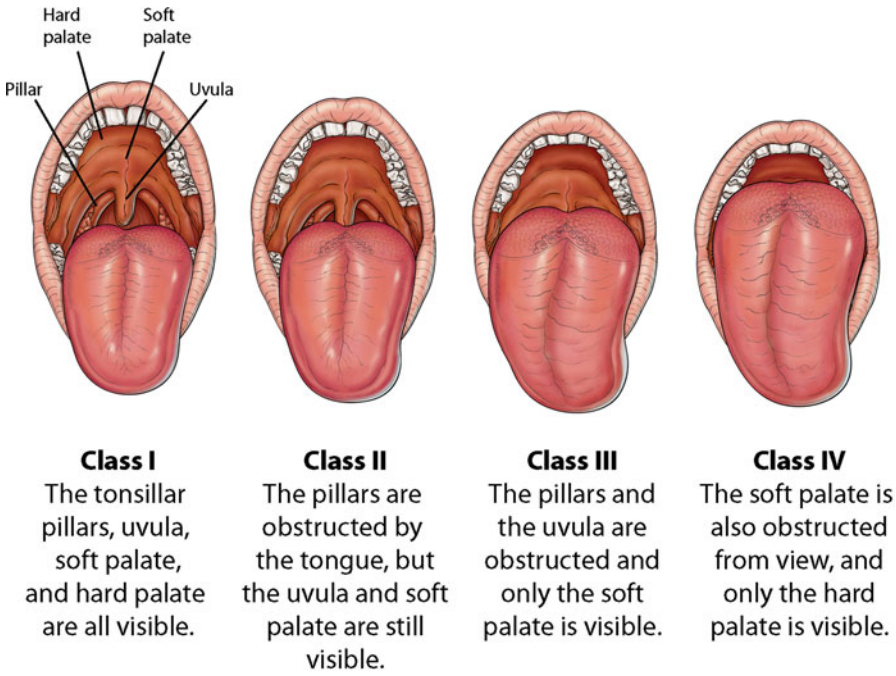

Fig. 8.1 Mallampati classification (Modified with permission from Finucane BT, Tsui BCH, Santora AH (eds). Principles of Airway Management, 4e (ISBN: 978-0-387-09557-8), New York, NY, Springer, 2011)

sequence of events in the OR if unexpected problems arise. The ASA taskforce has defined a well-outlined algorithm for managing these cases (Fig. 8.2). Prior to bringing a patient to the OR, one should consider four basic problems that may arise. These include difficulty with any of the following: ventilation, intubation, patient cooperation, or tracheostomy. The first management decision is whether the airway will primarily be secured by noninvasive techniques or through primary invasive surgical airway access. Once the decision is made to proceed with a noninvasive method, you will need to decide on whether to proceed with an awake intubation or intubation attempt after induction of anesthesia. This will primarily be based on your initial assessment as outlined above. Throughout the algorithm, various rescue methods exist to assist with securing a means of ventilation. Alternative noninvasive approaches include different laryngoscope blades, a laryngeal mask airway (LMA), fiber-optic technique, or esophageal-tracheal Combitube. Transtracheal jet ventilation is usually reserved as a last resort in patients that cannot be managed with noninvasive means and in whom emergent surgical access is contraindicated due to patient factors (anatomy, young age). Videolaryngoscopy is an emerging and useful technology that merits further study.

Different variations of surgical airway access exist and should be applied to the appropriate clinical circumstance. The choice of technique will again depend on patient factors (clinical status, neck anatomy), clinical experience, and setting (operating room, bedside, ER, etc.). A *cricothyrotomy* is the most accepted means of achieving rapid emergent airway access in a patient with no noninvasive options. This surgical technique is not a long-term solution to managing a patient's airway. Ultimately, a tube left this high and close to the glottis for an extended period of time would lead to subglottic (laryngotracheal) stenosis. It should be converted to a formal tracheostomy within 24–72 h after initial placement. For this reason, a cricothyrotomy is only preferred in emergent circumstances where rapid airway access is required, and further delay to prepare for a tracheostomy would lead to an adverse outcome.

The cricothyroid membrane is a horizontally oriented structure that lies between the inferior border of the thyroid cartilage and superior border of the cricoid cartilage. It lies superior to the trachea and closer to the skin surface with no intervening major structures such as the thyroid isthmus. A rapid wide transverse incision can be made over these landmarks and down through the membrane. Blunt hemostat dissection to dilate the opening in a superior-inferior direction will allow access to cannulate with an endotracheal or tracheostomy tube. Formal cricothyrotomy kits are typically available in most emergency rooms and crash carts.

An *emergent tracheostomy* can be attempted in the appropriate clinical circumstances. It is typically done at the bedside in the ICU, ER, or operating room. The patient should have somewhat favorable anatomy and appropriate

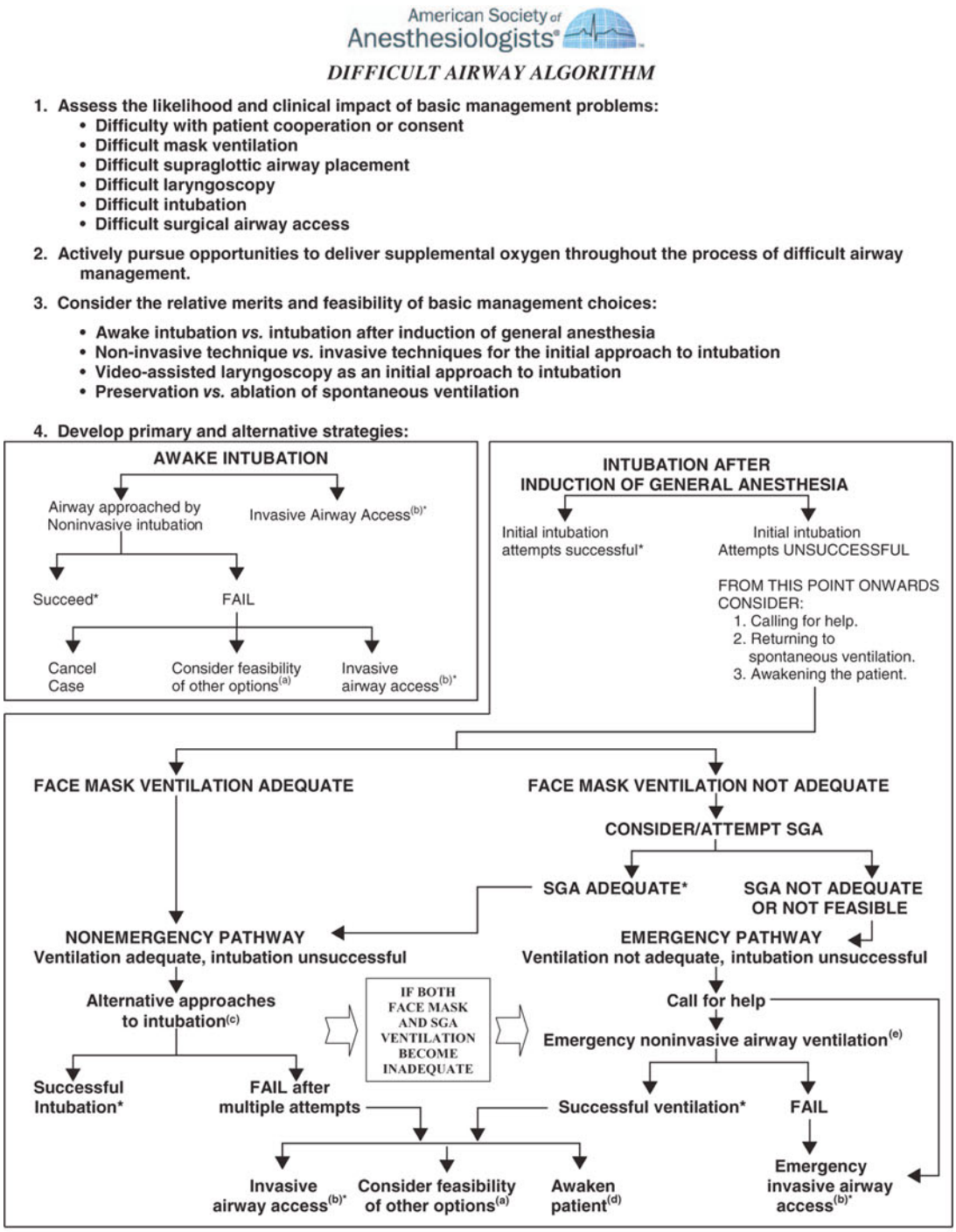

FIG. 8.2 Algorithm of difficult airway (Reprinted with permission from Apfelbaum JL, Hagberg CA, Caplan RA, Blitt CD, Connis RT, Nickinovich DG, et al.; American Society of Anesthesiologists Task Force on Management of the Difficult Airway. Practice Guidelines for Management of the Difficult Airway: An Updated Report by the American Society of Anesthesiologists Task Force on Management of the Difficult Airway. Anesthesiology. 2013 Feb;118(2):251–70)

equipment needs to be available (suction, lighting, instruments). This should only be attempted by a trained surgeon, and it is preferable to have a surgical assistant to help suction and retract if necessary. The cricoid cartilage is palpated and grasped with the left hand. A rapid vertical midline incision through the skin and subcutaneous tissue is made to avoid the anterior jugular veins, which can bleed quite profusely if encountered in the dissection. The strap muscles are bluntly dissected in the midline and retracted laterally, which should expose the thyroid isthmus. It will need to be retracted inferiorly. The thyroid is quite vascular and will bleed significantly if divided without electrocautery or suture ligature. The cricoid cartilage is grasped with a cricoid hook if available and the trachea incised in a vertical fashion. At this point, the field may be quite obscured and filled with blood requiring constant suction from your assistant. The wound should then be packed with gauze to give hemostasis, the patient stabilized, and then moved to the OR – if not already in that setting for control of bleeding and securing the tracheostomy tube.

An *urgent tracheostomy* is a more common scenario than the above two situations. These are patients for whom it is felt that airway compromise could be imminent and noninvasive attempts to secure the airway will have a high likelihood of failure. The presence of upper airway pathology (angioedema, tumor, infection) is typically present. Patients should be moved rapidly to the OR where a controlled sterile surgical setup is prepared in advance. These patients may be approached with a "double setup" where the anesthesia team will first attempt a noninvasive means to secure the airway, while the surgical team has the neck prepped and injected with local anesthetic. An *elective tracheostomy* is used in a prophylactic manner and is typically reserved for patients with head and neck cancer requiring further treatment with surgery, radiation, or both. These patients may have a prolonged course of upper airway swelling.

9
Penetrating Neck Trauma

Jennifer Rickard

Introduction

The workup and management of penetrating neck injuries has changed dramatically. As early as the 1500s, ligation of bleeding vessels was practiced for penetrating neck injuries. This resulted in increased survival, but increased morbidity due to neurovascular deficits. From the Civil War through WWII, most penetrating injuries were managed nonoperatively. This changed after WWII when concern for missed vascular injuries prompted many surgeons to perform immediate surgical exploration for all Zone II injuries. Since the 1990s, improvements in diagnostic imaging have allowed selective neck exploration based on physical exam and the results of imaging studies.

Mortality from penetrating neck injuries ranges from 2 to 10 %. Up to 15–20 % of patients have an arterial injury, and tracheolaryngeal injuries occur in 1–7 % of patients. Esophageal injuries are found in 0.9–6.6 % with a mortality rate up to 20 %. The mechanism of injury is important when considering the management of penetrating injuries. Gunshot wounds have a higher incidence of significant injury and therefore require surgery more frequently. A stab wound tract can be difficult to delineate, and the extent of the injury may, therefore, be underestimated.

Initial Evaluation

The evaluation of penetrating neck injuries begins with the basic ABCs per the American College of Surgeons Advanced Trauma Life Support (ATLS) guidelines. As with all patients, one must control the airway and assess breathing and circulation. If the patient requires an artificial airway, a surgical tray should be available because neck hematomas and tracheal injuries may complicate placement of an endotracheal tube.

Unstable Patient

Unstable patients should be taken directly to the operating room. This includes patients with an expanding hematoma, hemorrhage, airway compromise, shock, or subcutaneous emphysema. Other signs that mandate immediate intervention include neurologic deficit, stridor, dysphagia, or an impaled object.

A secondary survey should be performed assessing for concomitant injuries. A chest radiograph may be necessary to detect associated thoracic pathology, and an AP neck radiograph will show possible retained projectiles.

Explore Wound

The wound is then explored to determine the depth of penetration. Any wounds that do not violate the platysma can be safely monitored. Those that do cross the platysma are managed according to their location in the neck (see below).

Location in Neck

The neck is divided into anterior and posterior triangles relative to the posterior border of the sternocleidomastoid.

Anterior Triangle

The anterior triangle is further divided into three zones. *Zone I* extends from the sternal notch and clavicles to the cricoid cartilage. *Zone II* extends from the cricoid cartilage to the angle of the mandible. *Zone III* is above the angle of the mandible.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_9, © Springer New York 2015

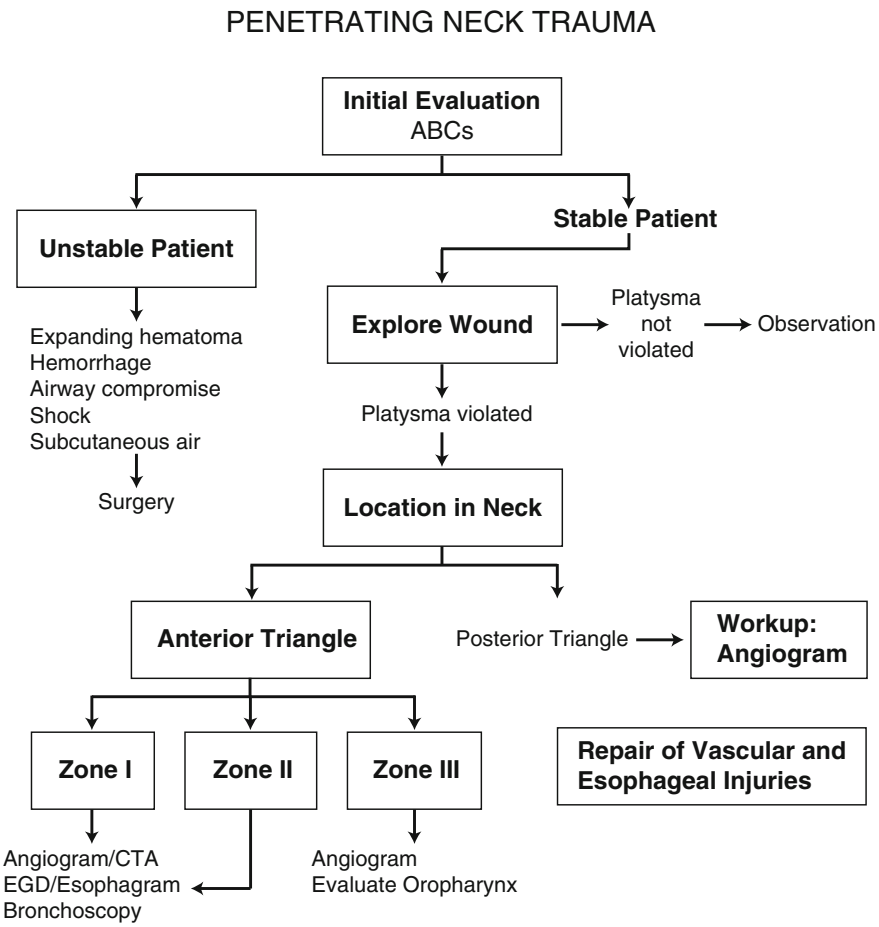

FIG. 9.1 Treatment algorithm for penetrating neck trauma

Based on the location of the wound, one can anticipate the structures that are at risk for injury. Zone I structures at risk include the aortic arch, innominate artery, brachiocephalic vein, subclavian artery and vein, common carotid artery, esophagus, trachea, thyroid, brachial plexus, lung apices, and thoracic duct. Zone II structures at risk include the common, internal, and external carotid arteries, jugular vein, pharynx, larynx, and esophagus. Zone II is the largest zone, most exposed, and therefore has the most injuries. It is also the easiest to explore. Zone III structures at risk include the internal carotid artery, external carotid artery branches, internal jugular vein, pharynx, and trunk of the facial nerve. Given the proximity to the skull base and overlying facial structures, it is a difficult region to explore.

Conventional angiography has been the gold standard for imaging vessels; however *CT angiography (CTA)* has shown comparative results in several studies and has the added benefit of showing the trajectory of the injury. In several studies CTA has a sensitivity and specificity of greater than 90 %. *Esophagoscopy (EGD)* and *contrast esophagography* are used to assess for injuries to the esophagus. When performed together, they have a sensitivity and specificity of 90–100 %. Airway injuries can be evaluated with *bronchoscopy and laryngoscopy*.

Zone I

Workup for a Zone I injury includes conventional angiography or CTA of the aortic arch and great vessels, esophagram/EGD, and bronchoscopy. Operative exploration may necessitate both a cervical and thoracic incision depending on the location and extent of injury.

Zone II

Workup of Zone II injuries includes conventional angiography/CTA, esophagram/EGD, and bronchoscopy. If there are signs of injury, proceed to the OR.

Zone III

Workup of Zone III includes conventional angiography/CTA and evaluation of the oropharynx. Given the location, these injuries are difficult to manage surgically. They may require a longitudinal neck incision with a clamp or Fogarty catheter passed distally for vascular control.

Workup: Angiogram

Posterior injuries are evaluated by conventional angiogram/CTA because the vertebral arteries course through the posterior triangle. All segments of the vertebral arteries are difficult to access surgically, and therefore endovascular therapy is the preferred treatment.

Repair of Vascular and Esophageal Injuries

Common and internal carotid injuries are best repaired surgically. This may include excision with interposition vein graft, primary repair, patch angioplasty, or transposition of the external carotid onto the internal carotid distal to the injury. Ligation of the common or internal carotid arteries carries a 30 % risk of stroke. In general, *the external carotid artery or internal jugular vein can be ligated without significant associated morbidity*. The trachea can be repaired primarily. The trachea is rarely injured but when it is, the complications are serious. The esophagus is repaired primarily and drained. Mortality is increased when the repair is delayed >24 h.

There are many vital structures contained within a compact space in the neck. One must be cognizant of these structures and carefully assess for all potential injuries and treat in a timely manner to minimize morbidity and mortality.

10
Penetrating Chest Trauma

James L. Lubawski Jr.

Introduction

The ABCs are the backbone of the trauma primary survey (Fig. 10.1). Nowhere is this more important than in the setting of penetrating chest trauma. The *A*irway (trachea), *B*reathing (lungs), and *C*irculation (heart and aorta) all have their most vital components within the chest.

Airway/Breathing

Tracheobronchial Injury

This unusual injury is often fatal in the field. Hemoptysis, subcutaneous emphysema, tension pneumothorax, or pneumothorax with a large, persistent air leak despite chest tube placement should all raise suspicion of this type of injury. Bronchoscopy is confirmatory. Intubation of the uninjured main stem bronchus should be attempted, if possible. However, accompanying injuries may make intubation extremely difficult, and immediate operative intervention should be undertaken to establish and repair the airway.

Open Pneumothorax

Also known as a sucking chest wound, an open pneumothorax is defined as an opening in the chest wall approximately two-thirds the diameter of the trachea. This allows for equilibration of chest and atmospheric pressures. Also, it allows for air to preferentially enter through the chest wound instead of the airway upon respiratory effort. This represents complete impairment of effective ventilation with resultant hypoxia and hypercarbia.

Treatment is an occlusive, sterile dressing applied to the chest wall defect. The dressing must be securely taped on only three sides. This allows for air to exit the thorax on exhalation, but prevents air entry on inhalation. Following this dressing, a chest tube is placed on the affected side, and definitive surgical closure is planned when the patient is stable. *Taping the dressing on four sides prevents air exit and results in tension pneumothorax.*

Tension Pneumothorax

Tension pneumothorax is a well-known entity largely because of its potential for early mortality and the ease with which it is fixed. It occurs when an air leak in the chest has no means of escape. This one-way valve effect allows pressure to increase inside the chest with each breath. The pressure can build within the chest to the point at which the mediastinum shifts. This shift can turn off venous return to the heart from the superior and inferior vena cavae leading to hemodynamic instability. Tension pneumothorax can be the primary presentation, or it can be a late finding complicating a simple pneumothorax if positive pressure ventilation is applied to a patient with a lung injury but no chest tube. Signs of tension pneumothorax include tracheal deviation, jugular venous distension, and absent breath sounds with hyperresonance on percussion. Initial relief of pressure is completed by the insertion of a large bore angiocatheter in the second intercostal space (found lateral to the sternum's angle of Louis). A rush of air from the catheter and hemodynamic improvement confirm the diagnosis. Formal tube thoracostomy is the next step for definitive treatment. *Tension pneumothorax should always be a clinical diagnosis. One should not wait for radiologic confirmation before treatment is undertaken.*

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_10, © Springer New York 2015

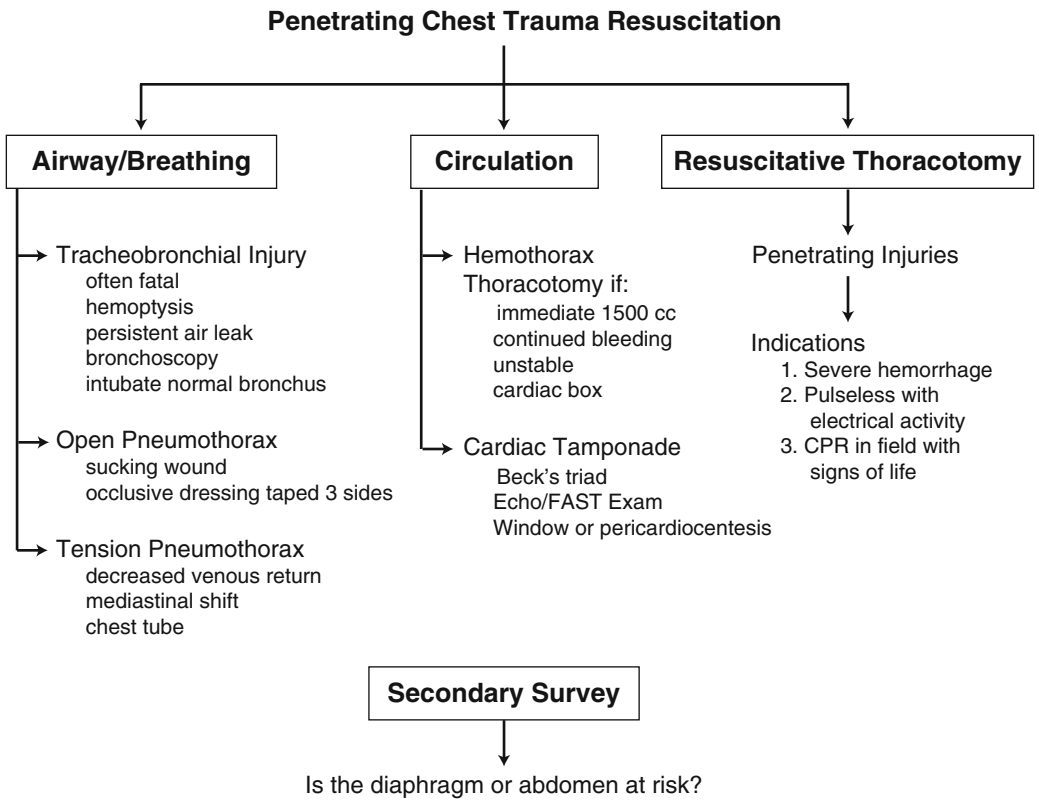

Fig. 10.1 Treatment algorithm for penetrating chest trauma

Circulation

Hemothorax

Hemothorax in penetrating chest wounds presents in a similar fashion to tension pneumothorax. In both, the patient is in shock with loss of breath sounds. However, if the chest is dull on percussion, hemothorax can be presumed. Chest tube placement and aggressive resuscitation with crystalloid and blood product infusion are the mainstays of treatment. *Urgent thoracotomy is warranted if 1,500 ml is immediately evacuated or if there is a high rate of continuing blood loss (200 ml/h × 4 h)*. However, hemodynamic instability or persistent need for blood transfusion may also warrant early surgical intervention. A penetrating wound to the "cardiac box" – an area bounded by the sternal notch superiorly, xiphoid process inferiorly, and laterally by the nipples (anterior) or scapula (posterior) – should raise suspicion of an injury to great vessels, hilar structures, or the heart. Urgent thoracotomy should be considered in these cases.

Cardiac Tamponade

A penetrating wound to the heart can result in cardiac tamponade. The pericardium is a relatively non-distensible fibrous sac. Addition of even small amounts of blood in a trauma can restrict right-sided heart filling and thus cardiac function. The classic clinical diagnosis of tamponade is by *Beck's Triad*: *decrease in arterial blood pressure, increase in jugular venous distension, and muffled heart sounds*. The diagnosis should be considered for any patient in hemorrhagic shock who does not respond to resuscitation. When tamponade is suspected, the diagnosis can be confirmed by echocardiogram or as part of a focused assessment with sonography for trauma (FAST) exam. In experienced hands, this method is 90 % accurate. Diagnosis and treatment can be completed concurrently with a pericardial window or, if surgical intervention is not feasible, pericardiocentesis.

Resuscitative Thoracotomy

The resuscitative or emergency thoracotomy is reserved for a relative few patients. First, only those who have penetrating thoracic injuries are candidates. Among those, the following scenarios warrant this potentially lifesaving procedure:

- Severe hemorrhage unresponsive to fluid resuscitation
- Pulseless with myocardial electrical activity
- CPR in the field with signs of life:
  - Signs of life include: reactive pupils, corneal or gag reflexes, and GCS >3.

Upon entry into the chest, a number of measures can be taken depending on the clinical situation:

- Pericardiotomy for release of tamponade
- Hemorrhage control
- Internal cardiac massage
- Aortic cross clamping

Secondary Survey

The careful examination of a patient with penetrating thoracic trauma during the secondary survey is important to find other less immediately threatening injuries. In addition to physical exam, upright chest X-ray, ABG, and ECG should all be conducted. Simple pneumothorax and hemothorax are found on chest X-ray and treated with tube thoracostomy. *Esophageal injury should be suspected in any transmediastinal injury*. This is best found by a combination of esophagoscopy and esophogram. Treatment consists of urgent primary repair and wide pleural drainage. Penetrating trauma to the thoracoabdominal area puts the diaphragm at risk for perforations. These injuries are often small and may take years to develop into larger holes through which abdominal contents may herniate. Diagnosis is difficult and may be made by X-ray (elevated diaphragm, bowel, or NG tube above the diaphragm), peritoneal lavage (RBC count >10 K, or lavage fluid in chest tube), or direct examination (laparoscopy, thoracoscopy). Treatment is direct repair. Thoracic duct injuries are found when milky chest tube drainage is seen. Confirmation is done by sending this fluid for triglycerides (>110 mg/dl) and protein (>3 g). Nonoperative treatment consists of low-fat diet and, if that fails, parenteral nutrition and octreotide. High-output leaks (>1,000 ml/day) usually require an operative intervention of direct ligation with or without talc pleurodesis.

11
Penetrating Abdominal Trauma

Aisha Shaheen and Marie Crandall

Introduction

The goal of managing patients with penetrating abdominal trauma is to quickly and efficiently assess injuries and establish treatment priorities (Fig. 11.1). The focus of the primary survey is resuscitation with continuous assessment of vital signs and control of hemorrhage. This is followed by a detailed secondary survey and initiation of definitive care; this is influenced by the hemodynamic stability of the patient as well as the location and mechanism of injury.

Primary Survey

The primary survey begins with the ABCs of trauma (*A*irway maintenance with cervical spine protection, *B*reathing and ventilation, and *C*irculation with hemorrhage control). Two large-bore (18 gauge or greater) peripheral intravenous lines are inserted, and a 2-l crystalloid bolus is administered. Life-threatening injuries should be identified, which requires complete disrobing of the patient. External hemorrhage should be identified and controlled with direct manual pressure.

Secondary Survey

The secondary survey involves a complete head-to-toe assessment of the patient. Continued management depends on (1) the hemodynamic stability of the patient, (2) the location of the injury, and (3) the mechanism of injury. Baseline laboratory studies include a complete blood count, electrolytes, coagulation studies, pregnancy test (for females), urinalysis, toxicology screen, and a type and cross matching. A nasogastric tube (NGT) – orogastric tube (OGT) if intubated – and Foley catheter should be inserted.

Assess Hemodynamic Stability

Hemodynamically Unstable *Patients*

Any patient with life-threatening hemorrhage or shock that is unresponsive to resuscitation efforts should be immediately taken to the operating room. Hemorrhagic shock can be classified into four categories based on the degree of total blood volume loss (Grade 1, up to15%; Grade 2, 15–30 %; Grade 3, 30–40 %; and Grade 4, >40 % loss of TBW) and accompanying signs and symptoms. Grades 3 and 4 hemorrhagic shock will manifest as profound hypotension and signs of compromised end-organ perfusion such as oliguria. Performing imaging studies on a hemodynamically unstable patient with this degree of blood loss serves no purpose.

Other indications for immediate surgical intervention include evisceration, free air, peritonitis, fascial disruption, retained stabbing implement, or gross blood per nasogastric tube or rectum.

Hemodynamically Stable *Patients*

Hemodynamically stable patients will have a Shock Index (SI = heart rate/systolic blood pressure) of 0.5–0.7 – as compared to SI >0.9 for hemodynamically unstable patients – and may undergo a more detailed evaluation with bedside diagnostic tests and imaging procedures in order to ensure there is no specific abdominal injury.

The choice of imaging studies is influenced by the location of the penetrating abdominal injury: whether it is anterior abdominal, thoracoabdominal, or in the back/flank region.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_11, © Springer New York 2015

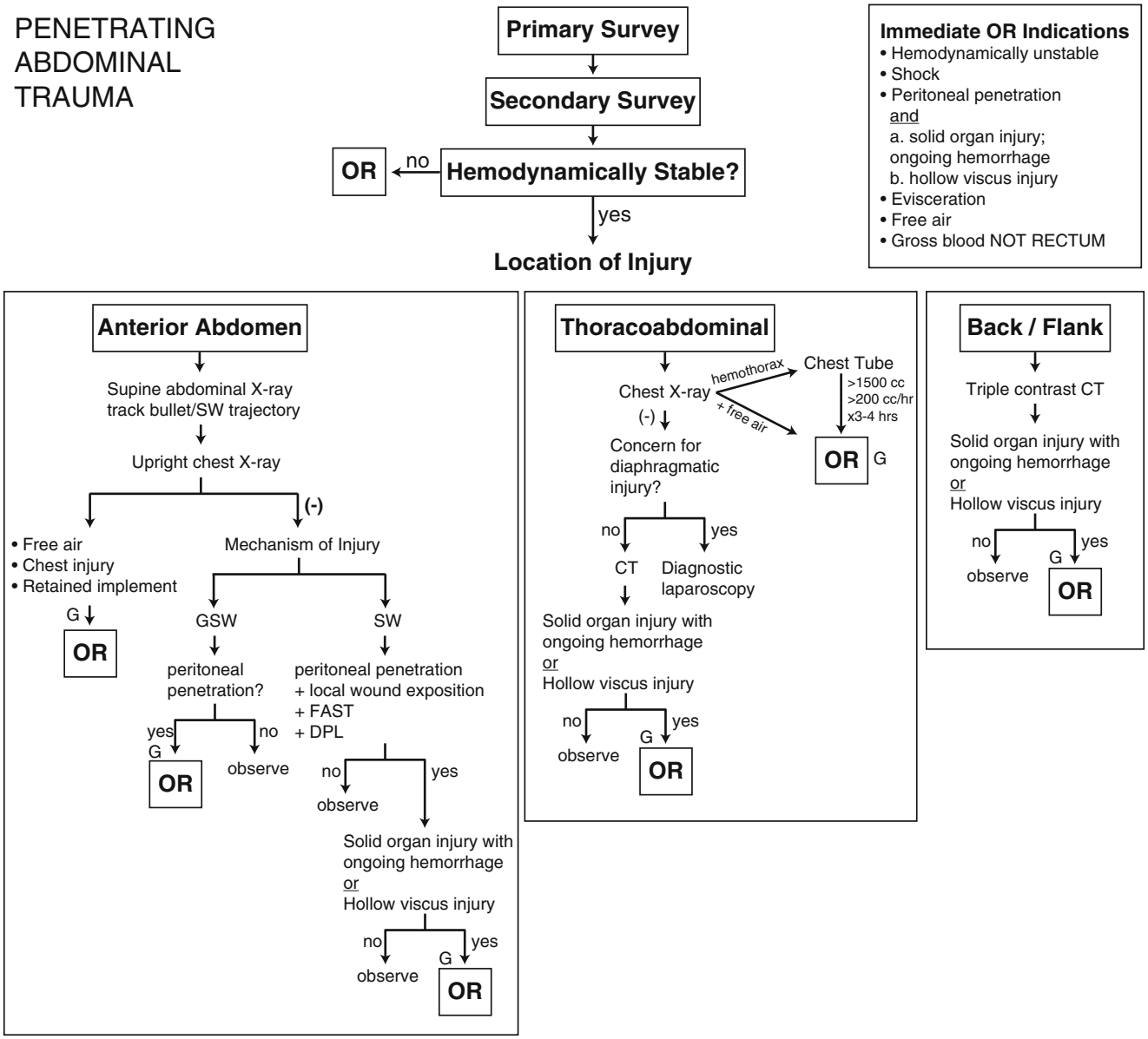

FIG. 11.1 Treatment algorithm for penetrating abdominal trauma. *OR* operating room, *GSW* gunshot wound, *SW* stab wound, *FAST* focused assessment with sonography for trauma, *DPL* diagnostic peritoneal lavage, *CT* computed tomography

Anterior Abdominal Injury

The anterior abdomen encompasses the space between the costal margins and the inguinal ligaments, anterior to the midaxillary line. Prior to obtaining any X-rays, all external entrance and exit wound sites should be marked with radio-opaque markers. An upright chest X-ray can be added to look for free air (evidence of a hollow viscus injury) or a concomitant chest injury.

For gunshot wounds (GSWs), special care must be taken to ensure all bullets are accounted for and their potential pathways of injury identified. In general, the number of holes added to the number of bullets should yield an even number. The treatment of GSWs to the anterior abdomen is most often an exploratory laparotomy even when the patient is hemodynamically stable or asymptomatic because of the high incidence of peritoneal penetration and associated visceral injury. For stab wounds, the likelihood of fascial penetration and associated peritoneal penetration is significantly lower compared to GSWs (50 % versus 85 %); consequently the management differs. Options to determine if surgical intervention is required include serial abdominal exams, local wound exploration, the focused abdominal sonography for trauma – or focused assessment with sonography for trauma (FAST) – exam, diagnostic peritoneal lavage (DPL), CT scans, or diagnostic laparoscopy.

If there is suspicion that the penetrating stab wound is superficial and does not violate the peritoneum, fascial integrity can be confirmed by local wound exploration. This requires adequate local anesthetic, good lighting, and a cooperative patient. Two diagnostic tests that can be used to rapidly detect hemoperitoneum in patients without obvious clinical manifestations of ongoing blood loss are the FAST scan and DPL.

FAST scan uses ultrasound technology to survey the pericardial sac and dependent regions of the abdomen for free fluid. The test is rapid and noninvasive; however, accuracy is operator dependant, and it cannot identify hollow viscus injuries.

DPL is a rapid, invasive test that can be used to identify intraperitoneal bleeding and hollow viscus injuries. The procedure involves aspirating intraperitoneal blood using a syringe. If no blood is aspirated, the peritoneal cavity is lavaged and the effluent evaluated for bile and bowel contents. A positive DPL includes aspiration of *gross blood, bile, or bowel contents* and mandates immediate laparotomy. DPL is primarily used for patients with no hemodynamic abnormalities when the FAST scan or computed tomography (CT) is not available. Of note, DPL is associated with a 5–12 % false-positive rate, which can result in a significant number of negative laparotomies.

CT scans are the gold standard for back/flank injuries and are increasingly being used for anterior abdominal stab wounds. CT scanning can identify tangential injuries, but lacks sensitivity for diaphragmatic injuries and can miss other occult injuries. Other limitations include the time required to complete the study, need for patient cooperation, and potential contrast allergy. CT scans can provide information about injuries that would be otherwise missed by physical exam, FAST scan, or peritoneal lavage.

Diagnostic laparoscopy can be used to rule out peritoneal penetration and to manage small splenic, liver, and diaphragm injuries. However, CT scans have largely replaced laparoscopy in evaluating penetrating abdominal injuries, except in circumstances concerning for diaphragmatic injury.

Regardless of the degree of suspicion for fascial disruption, retained stabbing implements should only be removed under direct vision in the operating room. To do otherwise may result in loss of vascular tamponade with subsequent massive, uncontrollable hemorrhage and ultimately death.

Thoracoabdominal Injury

The thoracoabdominal region includes the area below the nipples/scapula and above the costal margin. The diaphragm is at risk with penetrating injuries to this area, particularly on the left side because the liver does not extend to protect it. Diagnostic options for asymptomatic, hemodynamically normal patients include serial chest X-rays, CT scan, or laparoscopy. Chest X-ray is relatively insensitive but may show hemidiaphragm elevation, “blunting” of the hemidiaphragm from the presence of fluid, hemothorax, or free intraperitoneal air.

Just as with anterior abdominal injuries, CT scan for thoracoabdominal injuries can provide information relative to specific organ injury and can also diagnose retroperitoneal injuries. CT scans are not particularly sensitive for detecting diaphragmatic injuries; if patients have a concerning trajectory, laparoscopy is a definitive study.

Back/Flank Injury

Bordered by the scapulae superiorly and the iliac crest inferiorly, posterior to the midaxillary line, penetrating injuries in the back flank region can affect structures in both the peritoneum and the retroperitoneum. Laparotomy is chosen for symptomatic patients. For patients without an urgent indication for laparotomy, *abdominal CT scan with IV and rectal contrast* is the test of choice. Eighty percent of hemodynamically stable patients with back and flank injuries will have no injuries that require surgical intervention.

Operating Room

Many penetrating abdominal injury patients will be brought to the operating room because of hemodynamic instability, presence of symptoms, or fascial penetration. The first priority should be control of hemorrhage. This is accomplished via midline incision followed by a rapid but thorough survey and packing of all abdominal quadrants. Once hemorrhage is controlled, focus should shift to controlling contamination from hollow viscus injuries. Repair or resection may be indicated. Once necessary repairs have been completed, the patient should be transferred to a monitored setting and assessed regularly for any change in clinical status.

12
Blunt Abdominal Trauma

Jamie J. Coleman

Introduction

The diagnosis and management of blunt abdominal trauma is complex (Fig. 12.1) and has changed significantly in the past 20 years. The spleen and liver are the two most commonly injured organs in blunt abdominal trauma, and the trend towards their nonoperative management has been successful. Injuries to the pancreas, intestinal mesentery, or hollow viscera are important since they can be missed on initial examination, leading to significant morbidity.

Initial Management

Primary Survey

In accordance with the American College of Surgeons Advanced Trauma Life Support (ATLS) guidelines, the initial management of every trauma patient begins with the primary survey, commonly referred to as the "ABCDEs." The patient's *A*irway is first evaluated to verify whether or not it is protected or intact; *B*reathing is assessed for bilateral air exchange; and *C*irculation status can be determined by palpating pulses and measuring blood pressure. The patient's neurological status or *D*isability is assessed, and the patient should be completely undressed (*E*xposure) for a complete physical examination.

Secondary Survey

The secondary survey follows the primary survey and resuscitation and consists of a head-to-toe examination of the patient. After this exam is completed, chest and pelvis radiographs should be obtained as needed. For patients with blunt abdominal trauma, the physical examination should include inspection for any external signs of trauma (i.e., seat belt sign, road rash, etc.) as well as palpation for tenderness, distention, and signs of peritonitis. If signs of obvious intra-abdominal injury such as evisceration or generalized peritonitis are present, the patient should be taken directly to the operating room for exploratory laparotomy.

Exam Is Unremarkable, Equivocal, or Unreliable

Reasons that the abdominal examination may be compromised in the injured patient include depressed Glasgow Coma Scale (GCS), intoxication with alcohol or illicit drugs, and distracting orthopaedic injuries in the trunk. A focused assessment for the sonographic examination of the trauma patient (FAST) should be performed when the physical examination is not reliable. A 3.5 MHz convex transducer is used to evaluate for fluid in the following order: subxiphoid area to evaluate the pericardial sac, Morison's pouch in the right upper quadrant of the abdomen, splenorenal fossa in the left upper quadrant of the abdomen, and, lastly, the suprapubic area to evaluate for any blood in the pelvis. If the FAST exam is equivocal, then a further diagnostic work-up should be performed.

Positive Fast

Patients who are hemodynamically unstable and have a positive FAST examination are taken to the operating room, while stable patients should undergo an abdominal CT scan with contrast to further determine the origin of the intra-abdominal fluid.

Negative Fast

If the FAST examination is negative and the patient has a reliable and reproducible abdominal examination without findings indicative of an injury, no further work-up for abdominal

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_12, © Springer New York 2015

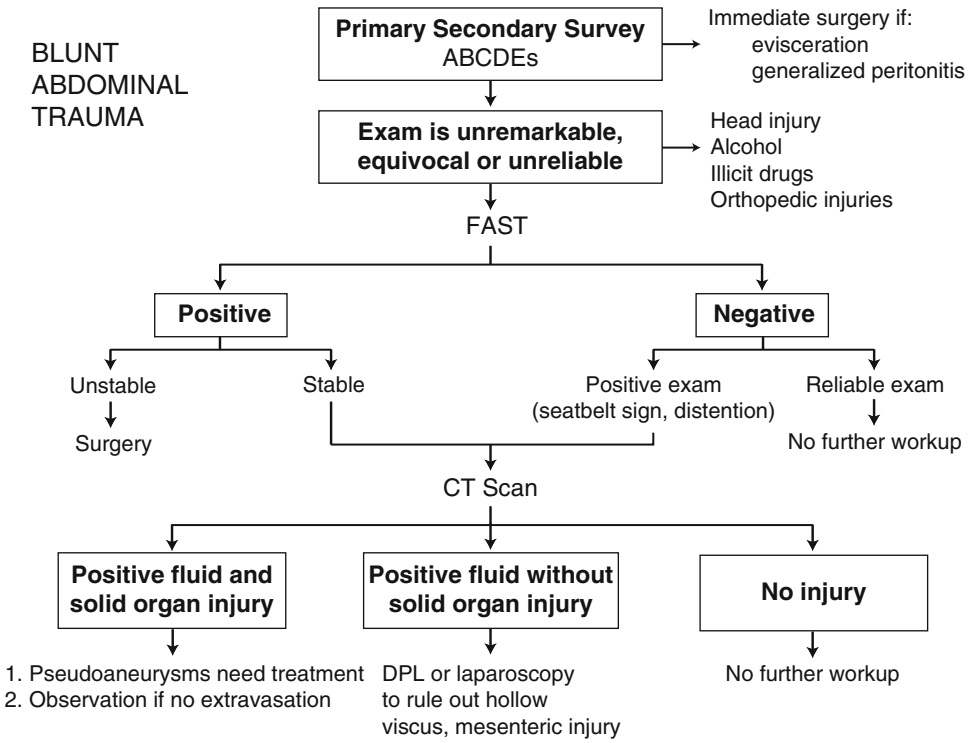

FIG. 12.1 Treatment algorithm for blunt abdominal trauma. *ABCDE* airway, breathing, circulation, disability, exposure, *CT* computed tomography, *DPL* diagnostic peritoneal lavage

injury is needed. Patients with physical findings concerning for abdominal trauma (i.e., seat belt sign, road rash, distention, pain) should have a CT scan of the abdomen with contrast.

Positive Fluid and Solid Organ Injury

Overall, approximately 80 % of patients with blunt hepatic injuries and 60 % of those with blunt splenic injuries can be managed nonoperatively. The presence of intra-abdominal fluid on the CT scan in the presence of an injury to a solid organ requires close examination of the injured organ in order to determine management. A CT scan that reveals active extravasation of contrast ("blush") or a pseudoaneurysm mandates further intervention by interventional radiology or a laparotomy. The choice of therapy is dependent upon hospital capability and the hemodynamic status of the patient. If the patient is hemodynamically stable and neither blush nor pseudoaneurysm is seen, the patient can be managed with observation in a monitored bed with serial abdominal exams and hemoglobin or hematocrit values. This is true no matter which solid organ or grade of injury is present. When these selection criteria are appropriately applied, the rates for successful nonoperative management of patients with blunt hepatic and splenic trauma are 98 % and 88–93 %, respectively. If nonoperative management is chosen, the patient is carefully monitored for changes in vital signs, hemoglobin and hematocrit values, or for new onset or increased abdominal pain and tenderness.

Positive Fluid Without Solid Organ Injury

If the CT scan shows intra-abdominal fluid without evidence of an injury to a solid organ, there is a concern about a subtle injury to the mesentery or to a hollow viscus. Further evaluation in such patients may include diagnostic peritoneal lavage (DPL), serial abdominal examinations if the patient is reliable, or diagnostic laparoscopy. The method chosen is often dependent upon the patient's alertness and cooperation, physician preference, and the resources available. A positive DPL in blunt abdominal trauma is defined as gross blood on aspiration, >100,000 RBC/mm^3 in at least 300 ml of returned fluid, >500 WBC/mm^3, amylase level>20 IU, and the presence of bile, feces, or food particles.

No Injury

If no evidence of injury is present on the CT scan, no further work-up is required to rule out intra-abdominal trauma.

13
Pelvic Fractures

Alexi Bloom

Pelvic Trauma

Pelvic fractures are typically associated with blunt trauma. Severity of pelvic trauma ranges from benign insignificant fractures to major vascular injuries leading to exsanguination. Initial management of pelvic injuries requires the determination of patient hemodynamic stability (Fig. 13.1). Hemorrhage control and volume resuscitation are critical in an unstable patient. These cases require a multidisciplinary approach to treatment; emergency medicine, trauma surgery, orthopedics, and interventional radiology must comanage these complex patients.

The pelvic ring is comprised of the sacrum, coccyx, ilium, ischium, and pubis. These bones are stabilized by a group of ligaments. Mainly, the sacroiliac, sacrospinous, and sacrotuberous ligaments provide support to the pelvic ring. Disruption of these ligaments leads to pelvic instability.

The pelvis is a highly vascularized space supplied by the internal iliac arteries. *The superior gluteal artery is the most common arterial branch to be injured by pelvic fractures.* Venous bleeding, however, is more common than arterial after pelvic injury. Attached to the anterior surface of the sacrum is an extensive venous plexus, which is often damaged after major pelvic fractures.

Fracture Classification

The Young-Burgess method of pelvic fracture classification first stratifies the injury by mechanism of trauma: lateral compression (LC), anteroposterior compression (APC), vertical shear (VS), and combinations of these injuries. Subclasses are further differentiated by severity of injury:

- LC fractures arise from lateral impacts commonly seen in motor vehicle accidents. They typically result in internal rotation of the hemipelvis. Type I fractures are stable and involve a sacral compression fracture on the side of impact and ipsilateral or bilateral pubic rami fractures. Type II are rotationally unstable but vertically stable fractures. These fractures involve an additional iliac wing fracture or posterior SI joint disruption.
- APC injuries are also often seen in motor vehicle collisions, but the hemipelvis displacement is demonstrated by external rotation. Type I fractures involve widening of the pubic symphysis <2.5 cm. Type II fractures involve pubic diastasis >2.5 cm along with anterior SI joint disruption, which contributes to the rotational instability of these injuries. Type III fractures involve additional disruption of the posterior SI joint, rendering these injuries rotationally and vertically unstable.
- VS injuries are often rotationally and vertically unstable. They result from falls leading to both anterior and posterior fractures.

Clinically, pelvic injuries are often combinations of these types of fractures. This stresses the importance of the history and physical exam for all patients with pelvic injuries. *Computed tomography (CT) has essentially replaced other diagnostic techniques in the characterization of pelvic fractures.* It also aids in the detection of hematomas and contrast extravasation. The focused abdominal sonography for trauma (FAST) exam is often performed to screen for intra-abdominal fluid, the presence or absence of which can prove useful in the evaluation of pelvic injuries.

Bleeding

Unstable fractures often lead to bleeding due to the movement of fractured elements and ongoing injury to surrounding vessels. *The goal is to prevent movement of the fractured elements and decrease the volume of the pelvis.* Stabilization can be achieved with pelvic binders. External fixation is a more permanent means of controlling bleeding. In its early stages, external fixation involves placement of pins in the superior iliac crest above the superior anterior iliac spine. Reduction of the fracture is then achieved by applying forces

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_13, © Springer New York 2015

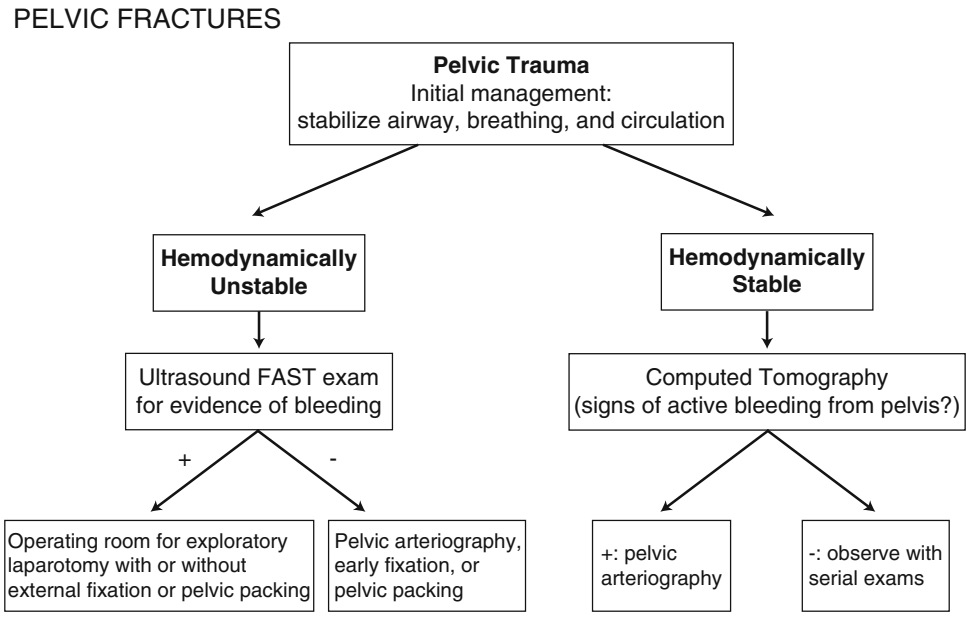

FIG. 13.1 Treatment algorithm for pelvic fractures. *FAST* focused abdominal sonography for trauma

opposite to those which caused the fracture. These pins remain for 6–12 weeks, after which a decision has to be made as to the long-term effectiveness of the external frame. *LC fractures, for example, often respond to external fixation. APC-III, most LC-II, and VS fractures, however, often require posterior stabilization by internal fixation.*

Access to angiographic embolization is critical in hemodynamically unstable patients. Interventional radiologists attempt to control bleeding by blocking the branches of the internal iliac arteries with agents such as gelatine sponge particles. These are temporary means of embolizing the internal iliac arteries, and they will recanalize within a few days. Collateral circulation ensures survival of pelvic tissues during this time. If angiography is not an option, then preperitoneal pelvic packing can be performed to emergently control bleeding.

14
Traumatic Hematuria

Stephen M. Larsen and Kalyan C. Latchamsetty

Introduction

Hematuria occurring in trauma patients is not a reliable predictor of urologic injury. The differential diagnosis is broad and spans the length of the genitourinary tract from the kidneys to the anterior urethra (Fig. 14.1). The mechanism of injury and location of trauma is important in rapidly directing appropriate diagnostic testing. The best indicator of significant urinary tract injury includes the presence of microscopic or gross hematuria with hypotension. However, the degree of hematuria and the severity of the urologic injury do not consistently correlate with each other as previous reports have demonstrated the absence of hematuria in 7 % of grade IV renal injuries. In general, genitourinary injuries are the least life threatening, but are most likely to affect long-term quality of life with respect to their complications.

History and Physical

The history and physical will usually steer the clinician toward an upper versus a lower source, given that upper and lower urinary tract injuries are almost never coincident (0.4 %). It is important to determine if a rapid deceleration event occurred (fall, high-speed motor vehicle accidents) or if there was a direct blow to the flank, pelvis, or external genitalia. For penetrating injuries, important information includes the size of the stabbing weapon and the type and caliber of the gun used. High-velocity projectiles have the potential for more extensive damage. When there is a bladder injury, conscious patients present with pronounced nonspecific symptoms such as suprapubic pain and inability to void. Urethral disruption is heralded by the triad of blood at the meatus, inability to urinate, and a palpably full bladder. Posterior urethral injuries typically occur with pelvic fractures, while anterior urethral injuries are associated with "straddle injuries" where the perineum and bulbar urethra are compressed between the pubis and a fixed object.

Physical examination is the basis for the initial assessment of each trauma patient, which may reveal obvious penetrating trauma from a stab wound or bullet entry or exit wound to the lower thoracic back, flanks, and upper abdomen. Flank ecchymoses and/or abrasions, fractured ribs, abdominal distension or tenderness, and palpable mass may indicate possible renal involvement. Physical signs of bladder injury include suprapubic tenderness, lower abdominal bruising, muscle guarding and rigidity, and diminished bowel sounds. Clinical signs of urethral injuries include blood at the meatus, gross hematuria, urinary retention, and other classic findings such as a "high-riding" prostate or a "butterfly" perineal hematoma. In severe trauma, Buck's fascia may be disrupted, resulting in blood and urinary extravasation into the scrotum. Women who develop proximal urethral avulsion injuries present with vulvar edema and blood at the vaginal introitus, thus indicating the need for careful vaginal examination in all female patients with pelvic fracture.

Lab Tests

The first aliquot of urine obtained either by catheterization or by voiding is used to determine the presence of hematuria. Later urine samples are often diluted by diuresis from resuscitation fluids, masking the presence of hematuria. Any degree of visible blood in the urine is regarded as gross hematuria. Microscopic hematuria can be detected by dipstick analysis or microanalysis. The dipstick method is rapid and has a sensitivity and specificity for microhematuria of more than 97 %. Microscopic hematuria in the trauma setting may be defined as greater than 5 red blood cells per high-power field (rbc/hpf).

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_14, © Springer New York 2015

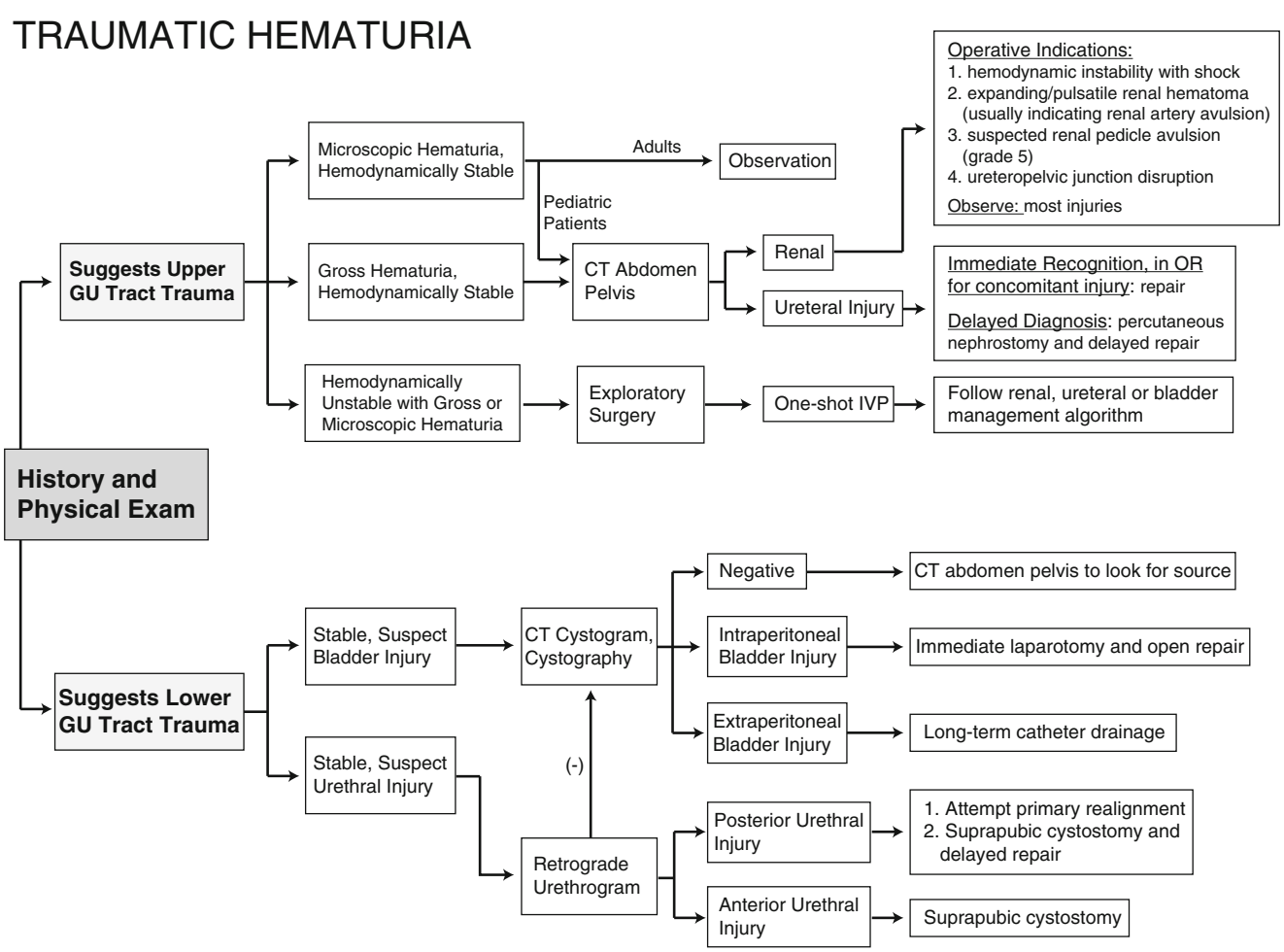

FIG. 14.1 Treatment algorithm for traumatic hematuria. *GU* genitourinary, *CT* computed tomography, *OR* operating room

Urologic Imaging

Kidney

The criteria for radiologic imaging include: (1) penetrating trauma with a likelihood of renal injury (abdomen, flank, or low chest) who are hemodynamically stable; (2) all blunt trauma with significant mechanism of injury, specifically rapid deceleration as would occur in a motor vehicle accident or a fall from heights; (3) all blunt trauma with gross hematuria; (4) all blunt trauma with hypotension defined as a systolic pressure of less than 90 mmHg at any time during evaluation and resuscitation; and (5) all pediatric patients with greater than 5 red blood cells (RBCs)/HPF.

Patients who are hemodynamically unstable after initial resuscitation require surgical intervention. Patients with microscopic hematuria without shock can be observed clinically without imaging studies. Contrast-enhanced computed tomography (CT) is the gold standard for genitourinary imaging. In addition to inspecting anatomic detail, one also looks for renal contrast extravasation and parenchymal/vascular injuries. Penetrating injuries with any degree of hematuria should be assessed with imaging studies if the patient is hemodynamically stable. The intraoperative "single-shot" intravenous pyelogram (IVP) can be considered when the surgeon encounters an unexpected retroperitoneal hematoma surrounding a kidney during abdominal exploration. The main purpose of the one-shot IVP is to assess the presence of a functioning contralateral kidney and to radiographically stage the injured side.

Ureter

Ureteral injuries after external violence are difficult to detect with the array of diagnostic tool. Preoperative urinalysis, CT scan, and intraoperative one-shot intravenous pyelogram (IVP) are nondiagnostic 33–100 % of the time. In the absence

of a better test, a one-shot IVP is still recommended together with intraoperative inspection to detect ureteral injuries in those patients who are undergoing surgery for concomitant injuries. The functional status of the contralateral system can be assessed with this as well. There are no absolute indications for CT in the management of ureteral injuries; thus they are obtained selectively. Ureteral injuries are suggested on CT by the absence of contrast in the ureter on delayed images, underscoring the importance of tracing both ureters throughout their entire course on CT scans. Delayed images must be obtained (5 to 20 min after contrast injection) to allow contrast material to extravasate from the injured collecting system, renal pelvis, or ureter. Retrograde ureterography is most commonly used to diagnose missed ureteral injuries because it allows the simultaneous placement of a ureteral stent if necessary.

Bladder

Imaging of the bladder is performed on the basis of clinical suspicion. A pelvic fracture occurring after blunt external trauma is an absolute indication for immediate cystography. Approximately 29 % of patients presenting with this combination of findings have bladder rupture. Relative indications for cystography after blunt trauma include gross hematuria without pelvic fracture and microhematuria with pelvic fracture. The diagnosis of bladder rupture is extremely low in these atypical groups (e.g., 0.6 % in patients with pelvic fracture and microhematuria), but the index of suspicion should be raised by associated clinical indicators of bladder injury. Conversely, penetrating injuries of the buttock, pelvis, or lower abdomen with *any* degree of hematuria warrant cystography. Retrograde or stress cystography is nearly 100 % accurate for bladder injury if performed appropriately. A dense flame-shaped collection of contrast suggests extraperitoneal extravasation versus intraperitoneal extravasation, which typically demonstrates contrast material outlining loops of bowel and/or the lower lateral portion of the peritoneal cavity. Since CT is now routinely used to assess trauma patients, concomitant CT cystography is now frequently selected as a more efficient means to assess the bladder. It is as accurate and reliable as plain film cystography and can easily delineate intraperitoneal and extraperitoneal bladder injuries. If blood is noted at the meatus or a catheter does not pass easily, retrograde urethrography should be performed first because urethral injuries occur in 10 to 29 % of patients with bladder rupture.

Urethra

Urethrography should be performed in cases of suspected urethral injury. When blood at the urethral meatus is discovered, an immediate retrograde urethrogram should be performed to rule out urethral injury. Direct inspection by urethroscopy is suggested in lieu of urethrography in females with suspected urethral injury.

Differential Diagnosis and Management

Kidney

Hemodynamic stability is the primary criterion for the management of renal injuries. Findings on CT that raise suspicion for major injury are: (1) medial hematoma (suggests vascular injury), (2) medial injury with urinary extravasation (suggests renal pelvis or ureteropelvic junction avulsion injury), and (3) lack of contrast enhancement of the parenchyma (suggests arterial injury).

The presence of concomitant injuries often influences the management of renal trauma. Absolute indications for surgery include: (1) hemodynamic instability with shock, (2) expanding/pulsatile renal hematoma (usually indicating renal artery avulsion), (3) suspected renal pedicle avulsion (grade 5), and (4) ureteropelvic junction disruption. Relative indications are now rare: urinary extravasation together with nonviable tissue, renal injury together with colon/pancreatic injury, and a delayed diagnosis of arterial injury (which most likely will need delayed nephrectomy).

Ureter

Absence of hematuria with ureteral injuries may result from an adynamic, partially transected ureter or a complete ureteral transection. Approximately 25 to 45 % of ureteral injuries do not demonstrate microscopic hematuria; therefore, the diagnosis of ureteral injuries is often delayed. Ureteral injuries represent less than 1 % of all genitourinary trauma and often occur in association with other visceral injuries, leading to a high mortality rate. Rapid deceleration injuries disrupt the ureter at specific points along its course, namely, the ureterovesical and, more commonly, the ureteropelvic junction (UPJ). UPJ disruption consequent to blunt trauma is often missed because the patients do not always exhibit hematuria. Isolated ureteral injuries are extremely uncommon. Intraoperative assessment of ureteral integrity is performed by occlusion of the ureter with simultaneous intravenous injection of indigo carmine or methylene blue. If extravasation is noted or ureteral injury is otherwise discovered, immediate repair is of critical importance. Surgical repair usually involves debriding devitalized tissue, performing a watertight anastomosis, and ureteral stenting and drainage. Upper ureteral injuries can usually be repaired with primary reanastomosis, while lower ureteral injuries (below pelvic vessels) usually

require reimplantation into the bladder. If ureteral injuries are discovered after more than 5 days, they are managed with temporary urinary diversion with a nephrostomy tube and Foley catheter.

Bladder

Most blunt bladder injuries are the result of rapid-deceleration motor vehicle collisions, but many also occur with falls, crush injuries, assault, and blows to the lower abdomen. Disruption of the bony pelvis tends to tear the bladder at its fascial attachments, but bone fragments can also directly lacerate the organ. Bladder injures that occur with blunt external trauma are rarely isolated injuries – 80 to 94 % of patients have associated nonurologic injuries. Immediate catheterization should be performed when blunt bladder rupture is suspected; the most reliable indicator is gross hematuria, which is present in nearly all cases. Other important causes of bladder rupture include penetrating trauma and iatrogenic surgical complications. Spontaneous rupture may occur in patients with a history of neuropathic disease, preexisting bladder disease, or prior urologic surgery.

The most common associated injury is pelvic fracture, which is associated with 83 to 95 % of bladder injuries. The usual treatment of uncomplicated extraperitoneal bladder ruptures, when conditions are ideal, is conservative management with urethral catheter drainage alone. Blunt extraperitoneal injuries with complicating factors warrant immediate open repair to prevent complications such as fistula, abscess, and prolonged leak. If a stable patient is undergoing exploratory laparotomy for other injuries or for internal fixation of pelvic fracture, it is prudent to surgically repair the extraperitoneal rupture at the same setting. *All penetrating or intraperitoneal injuries resulting from external trauma should be managed by immediate operative repair.*

Urethra

Urethral disruption injuries typically occur in conjunction with multisystem trauma from vehicular accidents, falls, or industrial accidents. Urethral injuries are classified as anterior or posterior. The following are critical points regarding anterior injuries:

1. They are usually isolated injuries.
2. The majority occur after straddle injury.
3. Initial management is with suprapubic cystostomy.

The following are important regarding posterior injuries:

1. They typically occur simultaneous with other injuries and pelvic fractures.
2. It is managed with immediate suprapubic tube placement.
3. Delayed reconstruction is advised for complete disruptions.

An attempt at primary realignment of a urethral injury with a urethral catheter is reasonable; prolonged or repeated attempts risk infection.

15
Extremity Fractures

Jonathan M. Frank

Initial Evaluation

The assessment of extremity fractures begins with an overall evaluation of the traumatic insults that a patient may have incurred (Fig. 15.1). Using the principles of ATLS (advanced trauma life support) training, the surgeon begins with a primary survey – the ABCDEs of trauma resuscitation. Upon secondary survey, a head-to-toe evaluation with appropriate interventions is performed. It is important to keep in mind that fractures will have bleeding that may possibly be life threatening. Obvious external hemorrhage is best treated with direct pressure. Internal bleeding is best controlled with immobilization of the extremity, which may be done with a backboard for critical injuries; otherwise a splint is preferred. Orthogonal radiographs are promptly obtained for any areas with gross deformity, tenderness, or concern for injury due to mechanism. Images of the joint above and below an affected bone are also obtained. A thorough neurovascular exam is performed, and an assessment of the status of the soft tissues is made. In cases of amputation, the amputated part is cleansed with a lactated Ringer solution and then wrapped in solution-soaked gauze. This is then placed in a labeled plastic bag that is further placed in an ice slurry, being careful that the part does not freeze. Instances of suspected vascular injury are investigated with angiography. Clinical findings that will raise the index of suspicion include knee dislocations or extremity injury with an ABI (ankle-brachial index) of <0.9, a cool pale hand or foot with slow capillary refill, or high-energy insult to a vulnerable area. Finally, as part of the tertiary survey, all fractures, including open fractures, are monitored for the development of a compartment syndrome.

Compartment syndrome is a rise in pressure within a closed fascial compartment that results in microvascular compromise with consequent impairment of myoneural function and eventual necrosis of soft tissues. Although the arterial pressure may be strong enough to deliver a pulse distally, the interstitial compartment pressure is greater than the capillary pressure, and resultant ischemia, edema, and eventual infarction will occur if left untreated. The classical 6 Ps of compartment syndrome are pain, pallor, poikilothermia, pulselessness, paresthesia, and paralysis. However, the most sensitive finding is pain with passive stretch of the muscles within a compartment. A high index of suspicion for an impending compartment syndrome should be entertained for any patient with a tense compartment, severe pain out of proportion to the injury, and/or pain with passive stretch. Emergent fasciotomy is the treatment for this condition. In cases where the clinical picture may not be so clear, such as an unconscious patient, compartment pressures can be measured. Pressures that are within 30 mmHg of the systemic diastolic pressure indicate a compartment syndrome. Prompt reduction and immobilization of fractures, prevention of hypotension, and frequent assessment play an integral role in avoiding of compartment syndrome and its devastating sequela.

Open Fractures

A fracture that communicates with an overlying break in the skin is considered open. Exploration of the wound in the emergency department is indicated only if surgical delay is expected. In such a scenario, the wound is copiously irrigated with sterile saline. Investigation of the wound is limited as repeated exposure to the outside environment increases the risk of wound contamination, resulting infection, and potential for further hemorrhage. Bleeding is controlled with direct pressure rather than tourniquet. Intravenous antibiotics and tetanus prophylaxis are initiated as soon as possible. Skin and soft-tissue damage is assessed, and a saline-soaked dressing is placed over the wound.

For fractures involving a joint with a nearby soft-tissue injury that has a question of communication with the fracture, if fluid is observed to extravasate from the soft-tissue wound, then this is an open fracture and treated as such. Sterile saline is injected into the joint. Fractures are anatomically reduced and splinted and any concern for vascular

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_15, © Springer New York 2015

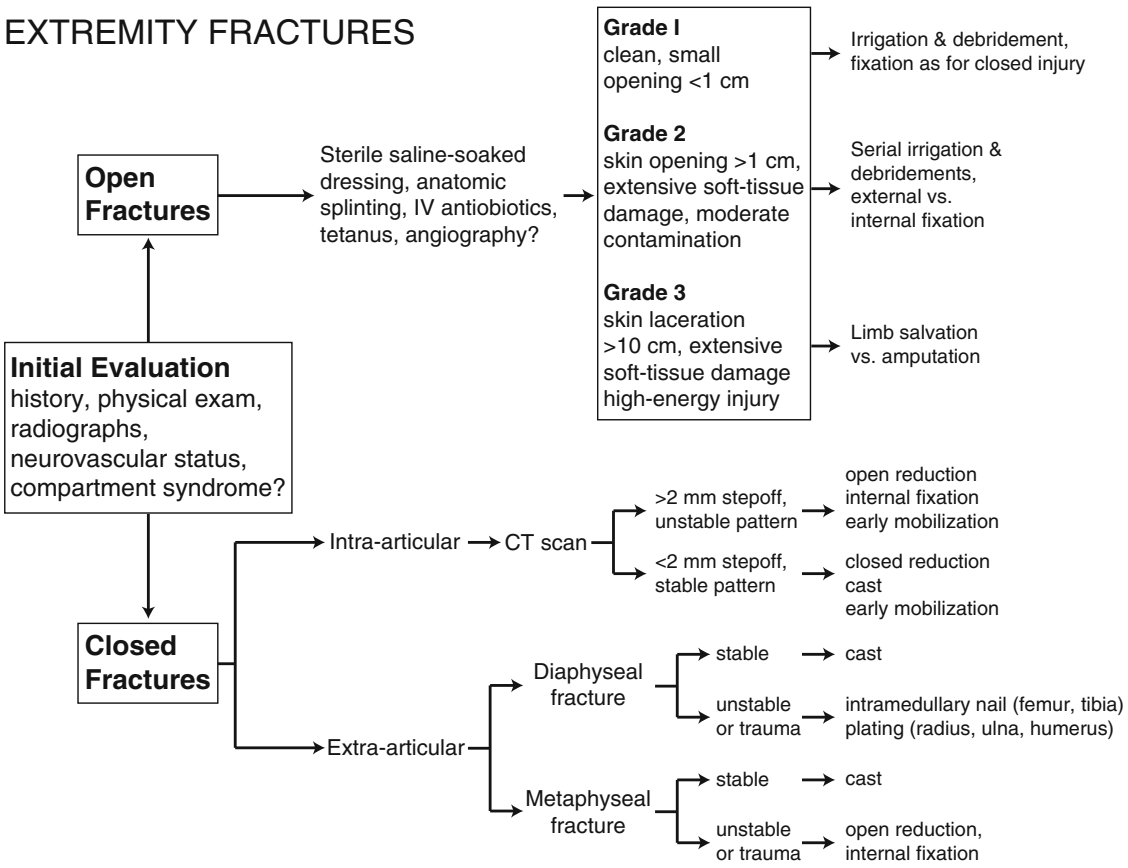

FIG. 15.1 Treatment algorithm for extremity fractures. *IV* intravenous, *CT* computed tomography

TABLE 15.1. Gustilo-Anderson classification of open fractures.

Gustilo-Anderson classification of open fractures			
Grade I		Skin opening <1 cm, minimal muscle contusion. Minimal contamination. Usually simple fracture pattern, i.e., simple transverse or short oblique	First-generation cephalosporin (or clindamycin if contraindication)
Grade II		Skin opening >1 cm, extensive soft-tissue damage but minimal-moderate crush component. Moderate contamination. Usually simple fracture pattern, i.e., simple transverse or short oblique with minimal comminution	First-generation cephalosporin (or clindamycin if contraindication)
Grade III		Skin laceration >10 cm, extensive soft-tissue damage. High-energy injury. Usually complex fracture pattern, i.e., comminuted and segmental	First-generation cephalosporin (or clindamycin if contraindication) + aminoglycoside (+PCN if farm contamination)
	A	Soft-tissue damage is extensive, but there is adequate bony coverage without the need for flap coverage	
	B	Soft-tissue damage is extensive with periosteal stripping and need for flap closure. Usually associated with considerable contamination	
	C	Vascular injury requiring repair; includes amputation	

injury addressed with angiographic studies. Operative fracture stabilization is important to prevent further soft-tissue damage and to provide maximum limb and patient mobility. The type of stabilization depends on the degree of soft-tissue injury and fracture location.

The choice of antibiotic depends on the grade of injury. The Gustilo-Anderson classification system grades open fractures based on the degree of soft-tissue injury, taking into account that a greater extent of soft-tissue injury will have a higher level of associated contamination (Table 15.1).

A Grade I fracture has a clean skin opening <1 cm with minimal muscle contusion. It is usually a simple fracture pattern such as a simple transverse or short oblique fracture. The wound is extended to expose and explore the injury zone and is sharply debrided. Intraoperative cultures are not necessary. Pulsatile lavage using saline with or

without antibiotics is used to irrigate the wound. The surgical portion of the wound is closed and soft-tissue coverage of the fracture obtained.

Grade II injuries have a skin opening >1 cm and extensive soft-tissue damage but a minimal-moderate crush component. They usually consist of a simple fracture pattern such as simple transverse or short oblique fracture with minimal comminution. Irrigation and debridement is performed as with Grade I injuries. An external fixator may need to be utilized for interim fracture stabilization to allow for the soft tissues to heal before pursuing definitive fixation.

Grade III injuries are high energy with a skin laceration >10 cm and extensive soft-tissue damage. They usually consist of a complex fracture pattern, i.e., comminuted and segmental. The decision between limb salvage and amputation is controversial. Several scoring systems – e.g., the MESS (Mangled Extremity Severity Score) – exist to aid the surgeon in this assessment. Indications for amputation include a nonviable limb with irreparable vascular injury, warm ischemia time >8 h, or an injury with insufficient remaining viable tissue. One must also consider the likely functionality of the limb should it be saved in contrast to the amount of potential function with amputation and prosthesis.

Closed Fractures

It is important to think about where the fracture occurred – that is, which bone is involved – not just where in the bone the fracture occurred. Several patient factors are considered when contemplating conservative versus operative treatment for fractures. These include age, activity, comorbidities, degree of osteoporosis and comminution, aspirations, and demands. Generally speaking, fractures in which articular congruity, length preservation, and axial and rotational alignment are essential for satisfactory function or outcome usually require surgery. If a satisfactory outcome requires anatomic restoration of the bone, those fractures will have to be treated surgically.

It is easiest to separate closed fractures into those that are intra-articular and those that are extra-articular. Intra-articular fractures have a stricter set of acceptable limits for closed reduction and conservative treatment in order to reduce the likelihood of posttraumatic arthritis and prevent arthrofibrosis. In general, if intra-articular step-off is >2 mm, open reduction and internal fixation should be performed. Computed tomography (CT) scan is useful in assisting with determining the amount of articular displacement. All intra-articular fracture-dislocations must be assessed for neurovascular injury. Again, the location and extremity of a fracture play an important role. Closed reduction and percutaneous fixation can be performed in instances where anatomic reduction is obtained with ligamentotaxis. Early and timely mobilization of the joint is important to maintaining mobility.

Diaphyseal fractures may be stable or unstable depending on the fracture pattern. Unstable fractures require surgical fixation. Lower extremity diaphyseal fractures are usually treated with intramedullary nailing because of the minimal disruption of the soft tissues and periosteum, as well as the faster return to full-weightbearing status. Open reduction and internal fixation is usually the preferred fixation for upper extremity fractures as these are non-weightbearing limbs. Stable long-bone fractures without significant varus-valgus or anterior-posterior angulation, rotational deformity, or shortening can be treated with casting and close serial radiographic observation.

Metaphyseal fractures are usually unstable, requiring open reduction and internal fixation. They are generally not amenable to intramedullary nailing due to the relatively wide medullary canal at the metaphysis and the resultant minimal bony purchase on the metaphyseal fragment. These fractures may be associated with neurovascular injury because of the tethering of these structures around joints, especially the elbow and knee.

16
Burns

Thomas Messer

Introduction

Burn injuries range from minor scalding injuries to large total body surface area (TBSA) flame burns resulting in systemic inflammatory response syndrome (SIRS), shock, sepsis, and multisystem organ failure. The initial approach to the burned patient is the same as for all injuries. Large TBSA burns carry a significant mortality risk throughout the hospital course, but failure to diagnose a solid organ injury or pneumothorax may be rapidly fatal. Every burn patient requires a full evaluation for trauma at the time of presentation (Fig. 16.1). Patient history is critical in this regard and should be obtained, when possible, prior to sedative administration or endotracheal intubation. The primary survey is focused on immediate threats to life. The secondary survey assesses the etiology, depth, and extent of burns and associated injuries. The American Burn Association (ABA) publishes guidelines regarding the transfer of patients to burn centers, available at www.ameriburn.org.

Primary Survey

The primary survey is the same as in other trauma patients, with a few additional concerns.

Airway

Airway is assessed during the initial examination of the patient. Indications for endotracheal intubation in all patients include depressed mental status, hypoxia, hypoventilation, or shock. The presence of inhalation injury should be considered in those patients exposed to fire, particularly in an enclosed space. Inhalation injury occurs due to the toxic effects of the products of combustion in smoke. Up to one-third of patients with major burn injury may suffer from concomitant smoke inhalation. Physical examination findings such as facial burns, singed nasal hairs, and carbonaceous sputum are frequently sought out, but are neither sensitive nor specific. Symptoms, including dyspnea, hoarseness, voice changes, odynophagia, and stridor, are more alarming and indicate the need to secure the airway. Endotracheal intubation should be performed urgently in these patients before upper airway edema worsens, making it difficult or impossible. Immediate surgical airway via cricothyroidotomy is indicated when endotracheal intubation fails due to upper airway obstruction.

Breathing

After establishing a patent airway, patients should be administered 100 % oxygen. Initial blood gas determinations include the measurement of carboxyhemoglobin (CO-Hb), as pulse oximetry will be falsely elevated in patients with carbon monoxide toxicity. One hundred percent oxygen should be administered for elevated levels until the CO-Hb is less than 10 %.

Circulation

Adults with greater than 15–20 % TBSA burned and children with greater than 10 % burns should receive intravenous crystalloid resuscitation. Burn shock is a unique phenomenon that requires large volume resuscitation due to the severe inflammatory response to injury. Two large-bore peripheral IVs in the bilateral antecubital fossae are preferred for resuscitation and may initially be placed through burned skin when necessary. Central venous catheters have higher complication rates, but will be needed when surface veins are unavailable or when invasive monitoring or vasopressors and inotropes are required.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_16, © Springer New York 2015

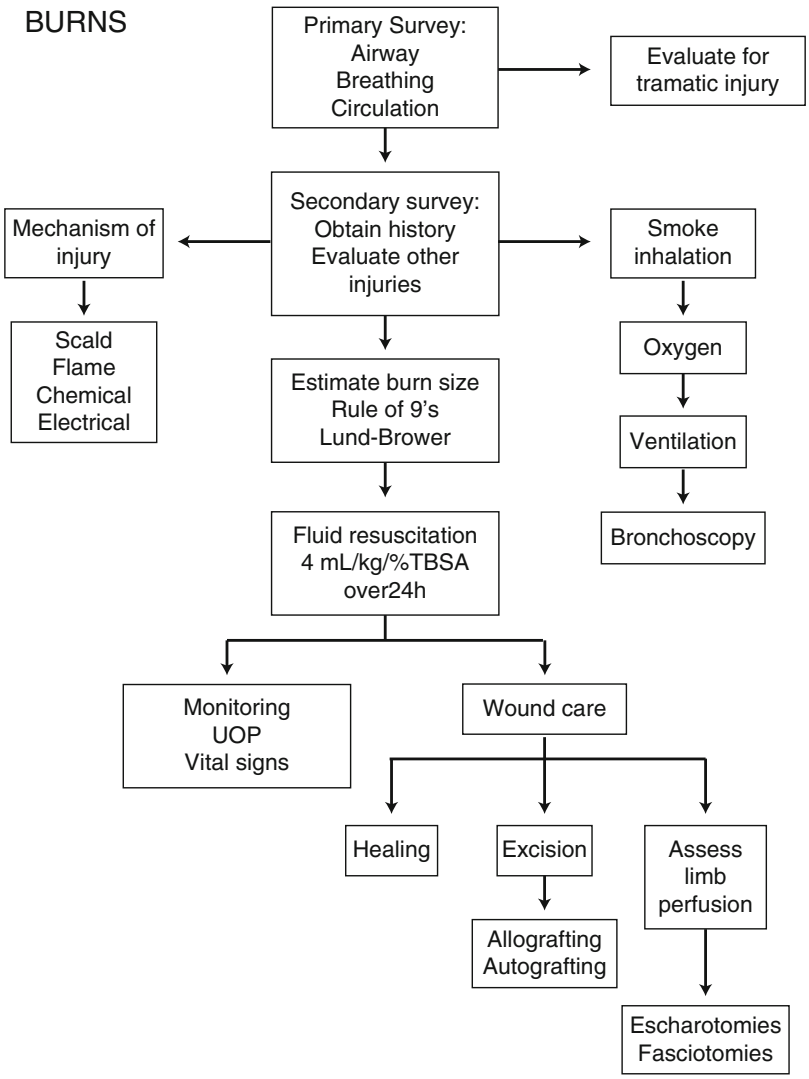

FIG. 16.1 Treatment algorithm for burns. *TBSA* total body surface area, *UOP* urine output

Secondary Survey

A full trauma evaluation should be undertaken as indicated by the patient history. Neurologic or torso injuries in particular may need to be diagnosed and managed prior to or simultaneously with burn care. Combined injuries are ideally managed at centers with both trauma and burn resources, or else initially at a trauma center until stabilized for transfer to a burn center.

The depth and extent of burn injuries is assessed during the secondary survey. Superficial (first degree) burns cause erythema and pain without epidermal loss. Superficial burns are common in sunburn or minor scald injuries, have little physiologic consequence, and are not included in calculations of TBSA burned. Partial-thickness (second-degree) burns involve the dermis and may be classified as superficial or deep. Epidermal loss exposes the injured underlying dermis. Superficial partial-thickness burns characteristically have epidermal blisters and exposed dermis that is pink, moist, blanching, and exquisitely tender. These burns usually heal within 14 days through epithelialization from remaining sweat glands and hair follicles with little or no scarring. Deep partial-thickness burns extend further into the dermis, resulting in more prolonged healing times (up to 2–4 weeks). Exposed dermis may be red, mottled, or pale in appearance. Scarring and contracture occur more frequently. Deep partial-thickness burns with healing time greater than 3 weeks may require skin grafting for optimal outcome. Full-thickness (third-degree) burns result in coagulation throughout the epidermal and dermal layers and, if not treated surgically, undergo contraction and epidermal ingrowth from the wound edges. Excision and split-thickness skin grafting are required to close full-thickness burns. Fourth-degree burns result from tissue loss down to muscle, tendon, or bone and are associated with prolonged contact with the heat source or high-voltage electrical injury. These wounds

present difficult reconstructive challenges, frequently requiring amputations. Early excision and autografting or allografting in large TBSA burns ameliorates the inflammatory response and the susceptibility to infection, which accounts for a majority of deaths in burn patients surviving initial resuscitation.

Estimation of the % TBSA burned may be first approximated using the "Rule of 9 s," which allocates 9 % to the head, 9 % to each upper extremity, 18 % (or two "9 s") to the anterior trunk, 18 % to the posterior trunk, 18 % to each lower extremity, and 1 % to the external genitalia. The Lund-Browder burn chart adjusts for the relative ratios of the head, thigh, and leg by age and is used to estimate burns more accurately upon arrival at the burn center.

Fluid resuscitation in burned patients is accomplished with continuous infusion of crystalloid (Ringer's lactate), titrated to maintain adequate hourly urine output (UOP) and other endpoints. There are many formulae for estimating the fluid resuscitation requirement in burn patients. The Parkland formula is widely used to predict the initial resuscitation and is straightforward to calculate. The Parkland formula estimates that patients suffering from major burns will require approximately 4 mL/kg/%TBSA burned of crystalloid over the first 24 h post burn. One half of the estimated volume is given as a continuous infusion divided over the first 8-h period, and the remainder given divided over the following 16 h. After initiation, volume infusion is titrated to maintain adequate hourly urine output (a surrogate for end-organ perfusion) or other secondary endpoints, with avoidance of bolus administration of fluids. In adults, adequate UOP is considered either 0.5 mL/kg/h or approximately 30–50 mL/h. In children the goal is 1 mL/kg/h UOP. Maintenance fluids must additionally be included for pediatric patients due to the higher proportional fluid losses and should be estimated as usual (e.g., the "4-2-1 rule") and given as additional dextrose-containing crystalloid. For large burns, albumin is given starting on the second postburn day once capillary leak has begun to resolve. Consideration may be given to earlier administration of albumin if a burn resuscitation is failing. When profound metabolic acidosis persists despite seemingly adequate resuscitation, occult injuries or cyanide toxicity should be sought out and treated. Nutritional support is critical to healing and survival. A nasogastric tube should be placed early in major burn patients and feedings instituted as soon as feasible as the patient is stabilized.

After the airway is secured and the patient stabilized, consideration should be given to bronchoscopy for diagnosis of suspected inhalation injury. Upper airway edema begins to subside over the days following admission, while lower airway injury due the toxic effects of inhaled smoke develops over several days postinjury. Supportive, lung-protective ventilation and aggressive pulmonary toilet are critical, and sequelae are common—including pneumonia, sepsis, airway obstruction, and acute respiratory distress syndrome (ARDS). The addition of inhalation injury to major burn injury increases overall mortality risk and may increase the volume of initial fluid resuscitation required by up to 25 %.

Hypothermia is common after major burn injury due to impaired thermoregulation. Prior to transfer to definitive burn care, patients should be kept warm and covered with clean, dry linens. Once admitted to the burn center, topical antimicrobials should be applied to burn wounds. Silver sulfadiazine is the most commonly used agent, has broad-spectrum activity, is soothing on application, and is widely available. Side effects may include leukopenia, allergic reactions, and irritation of eyes or mucous membranes. Mafenide acetate is also widely available, has a good spectrum of activity, and has the benefit of good eschar penetration. It is frequently recommended for severely burned ears to prevent suppurative chondritis. Mafenide acetate, however, is painful on application, may cause allergic reactions, and may cause metabolic acidosis when used in large quantities. Triple antibiotic ointment, with or without petrolatum gauze, may be used on facial burns or superficial partial-thickness burns to avoid irritation to eyes.

Examination of the extremities is important to assess the adequacy of distal perfusion. Circumferential full-thickness burns of the extremities may result in a tourniquet effect as progressive sub-eschar edema develops. Assessment of distal pulses may be aided by Doppler ultrasound, particularly when edema and burn shock make palpation unreliable. Serial examination of circumferential limb burns should be documented. Loss of pulses or signals, neurologic symptoms, and likelihood of progression of edema (as in the case of very large TBSA burns with multiple circumferentially burned limbs) are indications to perform escharotomies. Medial and lateral axial incisions are made, preferably with electrocautery, along the involved limbs through dermis into subcutaneous fat and extending the entire length of the burns onto healthy skin. Bilateral axillary incisions (may be connected by a transverse incision inferior to the pectoralis in an "H incision") are made in the case of circumferential torso burns if the patient becomes difficult to ventilate.

Part III
Vascular Problems

17
Chronic Lower Extremity Ischemia

Sherry Cavanagh and Chad E. Jacobs

History

Patients at most risk for this disease are those who have a smoking history (past or current), as well as other atherosclerotic disease processes such as diabetes, hypertension, and hyperlipidemia. The strongest correlation is seen with tobacco use; severe disease is rarely seen in the non-tobacco-using patient. Careful history taking is necessary to assess for risk factors and delineate arterial insufficiency from other etiologies that may mimic symptoms and findings. These include peripheral neuropathy, spinal stenosis, radiculopathies, arthritis, and rheumatologic diseases, or a combination thereof. Important information can be elicited that will guide decision making and clinical investigation (Fig. 17.1).

Functional Limb Ischemia

The hallmark of *functional limb ischemia* is intermittent claudication. Patients consistently experience pain in a functional muscle unit (i.e., calf) after a certain distance or time of activity. This is often quantified by the number of city blocks a patient can walk before having to rest to allow the pain to resolve. Claudication is relieved by a period of rest; some patients will describe having to massage the area as well. It is important to distinguish this symptom from muscle cramps (does not occur in claudication); spinal stenosis, where the patient may lean forward in a sitting position to alleviate the symptoms; or "shooting" pain across muscle units, which could indicate a neurologic pathology.

Critical Limb Ischemia

Rest pain is the key indicator of *critical limb ischemia*. In addition, tissue loss in the form of ulcerations or gangrene of the foot or toes is also diagnostic of critical ischemia. While claudication affects a muscle group, rest pain typically occurs in the toes or distal foot. This often occurs at night while the limb is elevated and blood flow to the distal extremity is not assisted by gravity. Key questioning of the patient surrounds their response to the pain and actions they may take to alleviate it, such as dangling the foot off the side of the bed or walking around in an attempt to increase circulation. Rest pain indicates inadequate perfusion to the tissues at baseline metabolism and if untreated will progress to tissue loss (ulcers, gangrene).

Physical Exam

The vascular exam assesses the entire cardiovascular system from head to toe. Auscultation over the carotid arteries is performed as is auscultation of the cardiac tones; the abdomen is auscultated over the renal arteries and the groin listening for bruits of the renal, iliac, and femoral vessels. The abdomen is palpated for any pulsatile masses, which may indicate the presence of an aortic aneurysm. The peripheral pulses are palpated including the radial, femoral, popliteal, dorsalis pedis, and posterior tibial arteries. If no pulse is palpable, vascular signals need to be assessed using a handheld Doppler. Other findings on physical exam to note are level of alopecia, ulcerations or wounds, skin temperature, venous stasis changes, gangrene, delayed capillary refill, and edema. Chronic arterial ischemia can lead to skin changes that appear shiny and tight or "skeletonized" due to atrophy of the skin and subcutaneous tissues.

Noninvasive Diagnostic Studies

The ankle-brachial index, or ABI, is a key test performed to assess the degree of arterial insufficiency. Stenotic lesions in an artery cause a decrease in the distal blood pressure, which is reflected in the ABI. The test can easily and rapidly be performed at the bedside with the use of a manual blood

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_17, © Springer New York 2015

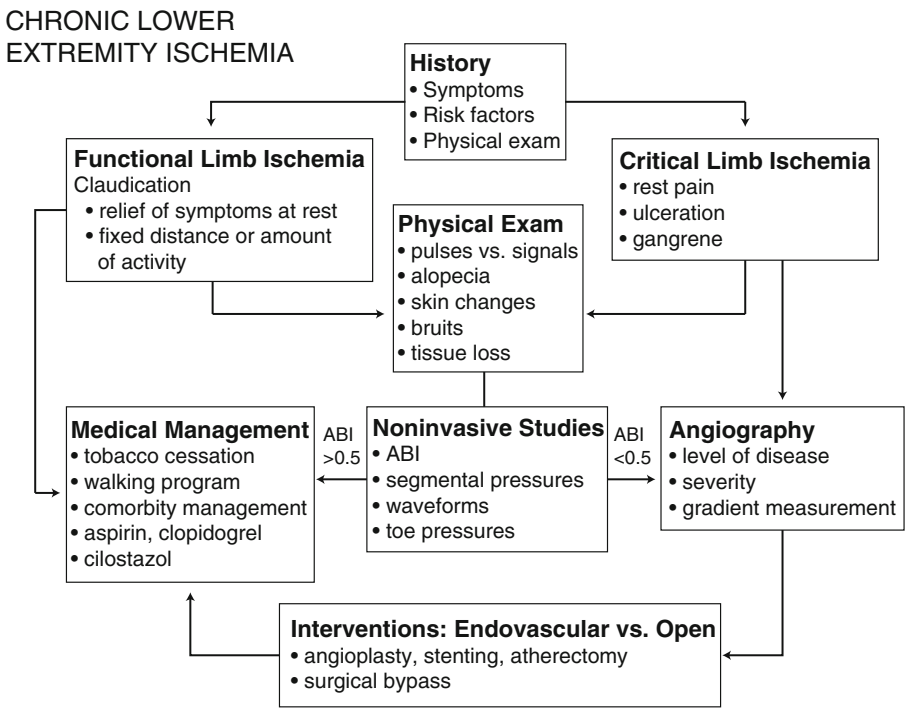

FIG. 17.1 Treatment algorithm for chronic lower extremity ischemia. *ABI* ankle-brachial index

pressure cuff and handheld Doppler. The right and left brachial systolic blood pressures are taken using a blood pressure cuff and Doppler, recording systolic values after inflating the cuff on the upper arm. The higher of the two brachial pressures will be used to calculate the ABI. Next, the cuff is placed above the ankle, and the dorsalis pedis (DP) and posterior tibial (PT) signals of each leg are assessed. The higher pressure of the two distal vessels of each leg will be used for the calculation. The ABI is calculated by dividing the higher of the two pedal (DP or PT) systolic values by the higher of the two brachial systolic values (left or right). A normal ABI is approximately 1.1, claudication is accurately predicted with values from 0.40 to 0.90, rest pain occurs in ranges of 0.20 to 0.50, and tissue loss usually occurs with values less than 0.20. While the ABI alone is useful for determining severity of arterial insufficiency, it does not provide information as to the location of the disease. The ABI may be normal in patients with symptoms consistent with intermittent claudication. In such cases, studies should be repeated with exercise testing, which may unmask lesions not detected at rest.

Formal ABI studies with waveform analysis and digital pressure measurements are often performed in the vascular laboratory. Segmental pressure measurement evaluates the systolic pressures at the thigh, above knee, below knee, ankle, and digital levels, recording Doppler signals at the ankle with at each level of cuff inflation; both waveforms and the pressures are noted in the results. This helps to identify the level of the lesion causing arterial insufficiency; a pressure drop of 15 mmHg or greater from one level to another is considered significant. Depending on the level of change seen in either pressures or waveform analysis, disease can be localized to the aortoiliac, femoral, or infrapopliteal level.

In patients with calcified vessels (often found in diabetics), the ABI and segmental pressures are falsely elevated or are not able to be obtained due to non-compressibility of the vessel. In these patients, toe pressures are very important in evaluating the patient, as the calcific disease tends not to affect the toe vessels. A toe pressure <30 mmHg indicates severe ischemia.

Duplex ultrasonography is another tool for evaluating patients with arterial insufficiency. It can be used to map the arterial tree and evaluate for stenosis and disease in a specified segment. It is very useful in the postoperative surveillance setting, particularly for evaluating patients who have undergone surgical bypass or percutaneous intervention. It also provides helpful diagnostic information in patients with renal insufficiency, who may be unable to undergo angiography for fear of renal contrast toxicity.

Angiography

While duplex ultrasonography and segmental pressures add much to the evaluation of limb ischemia, *angiography is the gold standard for assessment and planning of vascular interventions*. It has now also become widely used at the time of endovascular interventions as a treatment modality. Prior to performing contrast angiography, the physician must assess

the patient for contrast allergy and renal function. Allergic reactions to contrast may be prevented by using pre-procedure protocols for steroid and diphenhydramine administration, which typically starts 24 h prior to the administration of contrast. If the patient has mild creatinine elevation, fluid hydration with bicarbonate with or without mucomyst therapy should be performed pre- and post-procedure. The latter has mixed results in the literature as to the effect, but does not put the patient at risk when given; hence, some continue to use this medication despite a lack of definitive data. If the patient has more severe kidney disease but is not yet on dialysis, carbon dioxide (CO_2) angiography without the use of iodinated contrast may be performed. The latter provides detailed anatomic information despite the use of an alternative contrast agent.

The standard evaluation begins with aortography with bilateral lower extremity runoff to assess both proximal and peripheral disease. A luminal narrowing greater than 50 % is considered hemodynamically significant. If in doubt, intra-arterial pressure gradients on each side of a stenosis may be measured with a gradient of 15 mmHg considered significant.

Interventions

There are two principles in evaluating the treatment of limb ischemia: *inflow* and *outflow*. Inflow is the amount of blood getting into the limb, and outflow refers to the amount of blood that reaches the distal portion of the extremity. One or both may be diseased and require intervention.

If claudication is debilitating or prevents the patient from participating in routine activities of daily living or working, intervention is indicated. Those with critical limb ischemia need immediate assessment and intervention for preservation of the limb as they are at high risk of amputation, which in turn affects long-term mortality.

Aortoiliac disease can be unilateral or bilateral and can manifest as stenoses or occlusions. Stenotic lesions may be treated with endovascular angioplasty and stenting if focal or short-segment disease is present. This technique often results in a significant increase in the blood flow to the extremity. If long segment or nearly occlusive disease is present, surgical bypass is more likely to be warranted. Complete occlusions often require treatment with surgical bypass, but endovascular recanalization is an option as well. Iliac occlusions may be treated percutaneously by stent placement. Surgical options include direct reconstruction via aortobifemoral bypass or iliofemoral bypass. Extra-anatomic bypass via cross-femoral or axillo-femoral artery bypass are options as well but typically have lower long-term patency rates. In patients with inflow disease and distal lesions, treatment may be a combination of open surgical revascularization and endovascular interventions.

Infrainguinal disease may occur in any vessel of the lower extremity. Disease in the superficial femoral artery disease is most often found at the adductor (Hunter's) canal. Short-segment, focal disease in the above knee arterial tree may be amenable to endovascular procedures including angioplasty, stenting, and/or atherectomy. The latter is a percutaneous technology that improves blood flow by removing atherosclerotic plaque with either a rotating burr, an excisional rotating blade, or laser energy. Once across a lesion, it may then also be treated with balloon angioplasty and stenting if needed. If endovascular therapy is not an option, surgical revascularization with bypass grafting is performed. The preferred conduit is always autologous saphenous vein, usually harvested from the ipsilateral limb if it is of adequate size (>3 mm). If vein size is inadequate, a synthetic conduit may be used (polytetrafluoroethylene [PTFE] or Dacron), as in the above knee position, both autogenous and synthetic conduit materials have been shown to have similar patency rates.

Disease at the knee and in the infrapopliteal region may be amenable to endovascular treatment, once again in short-segment, focal, or multifocal disease. The endovascular procedures described above (angioplasty, stenting, and/or atherectomy) also apply to these territories. Stent placement at the knee is avoided out of concern for fracturing of the device with repeated bending of the joint.

The preferred conduit for revascularization to the infrapopliteal vessels remains autogenous saphenous vein. Configurations include either reversed saphenous vein graft (RSVG), orthograde saphenous vein, or in situ bypass. Below the knee, patency rates are significantly higher with vein (68–85 %) than for synthetic conduit (34–56 %). If no autogenous conduit is present, however, distal bypass with synthetic conduit can still be performed. Several technical variants exist for modifying the prosthetic distal anastomosis to increase patency, including vein patches and cuffs, or synthetic grafts with built-in expanded hoods.

Medical Management

Patients with functional limb ischemia and low risk of limb loss are first treated with medical management. *Of utmost importance is tobacco cessation in all groups*. Control of medical comorbidities such as diabetes, hyperlipidemia, and hypertension is imperative. *Regular exercise programs are recommended* as they promote formation of collateral vessels, improving blood flow and symptoms over time. For functional ischemia, medical management provides a reasonable outcome as only 5–7 % will eventually progress to require an amputation of the affected leg. Claudication patients are prescribed *cilostazol*, a phosphodiesterase inhibitor that causes vasodilation and inhibition of platelet aggregation, smooth muscle proliferation, and improvement in lipid profile. Cilostazol has been definitively shown to improve walking distance over placebo in claudicants, but must be avoided in patients with congestive heart failure.

Pentoxifylline, which decreases blood viscosity and platelet aggregation, may be prescribed in such patients, although it is not significantly better than placebo in controlled trials. All patients with peripheral arterial insufficiency of any severity are recommended to be on *aspirin* for its antiplatelet effect, as it not only lowers the risk of future limb surgery but also that of stroke or myocardial infarction. In addition, *clopidogrel* has demonstrated a relative risk reduction of cardiovascular events in patients with arterial insufficiency and may be prescribed if no contraindications are present.

18
Algorithmic Approach to the Acute Cold Leg

Benjamin B. Lind

Background

Emergent surgical consultation may be requested for the evaluation of the cold lower extremity. Timely recognition, diagnosis, and intervention are crucial for successful limb salvage. Permanent tissue damage can occur in approximately 6 h in the completely ischemic limb. Etiologies of ischemia include: embolism from a proximal source, such as intracardiac thrombus in the patient with arrhythmia; acute thrombosis of chronic, localized disease; arterial dissection; occlusion of preexisting bypass graft; and thrombosis due to hypercoagulable state. Although various etiologies may be responsible, the underlying tenets of diagnosis and treatment are constant (Fig. 18.1).

Diagnosis

History

A careful history will greatly aid the diagnosis of the acutely ischemic limb. Pain in the limb is almost universal and is usually the first symptom, as nerves are very sensitive to, and intolerant of, ischemia. Patients can often describe the exact moment of onset, as well as the inability to obtain relief. They may find some relief keeping the limb in a dependent position.

Recent procedures or medical events should be elucidated. Recent catheterization via the groin may suggest femoral or iliac artery trauma. Recent myocardial infarction or new onset arrhythmia may suggest a cardiac embolic source.

Pertinent medical history may include: cardiac arrhythmia; history of prior embolic or thrombotic events, including deep venous thrombosis; hypercoagulable disorder; collagen vascular disease; cardiac structural disease; aortic aneurysm or dissection; atherosclerotic disease; and preexisting claudication or rest pain.

Patients should be queried about medications and compliance with anticoagulant regimens. Pertinent social history should be gathered, including tobacco use. Prior vascular interventions, such as bypass grafting, as well as angioplasty and stenting should also be investigated.

Physical

Physical examination focusing on the cardiovascular system is performed. A complete pulse examination is obtained and is documented as "palpable," "signal," or "absent." The contralateral limb can often serve as a control, because chronic occlusive disease is often symmetric and acute disease usually affects one limb. Be aware that palpation of a pulse can be misleading because the examiner may be feeling his own pulse. Likewise, auscultation of a Doppler signal can mistakenly identify a phasic, venous signal for a low-intensity arterial signal. For these reason, a bedside ankle-brachial (or wrist-brachial) index should be obtained for a more objective assessment of limb perfusion.

Motor function, neurologic function, and skin status and integrity should be documented. The "rule of Ps" is a useful mnemonic for guiding the examination: pain, pallor, pulse deficit, paresthesia, and poikilothermy.

Physical findings can be used to classify acute limb ischemia (Table 18.1).

Decision Making

After limb viability and the ischemic threat to viability are determined, further workup and intervention are planned. Patients with a nonviable limb are offered amputation.

The patient is anticoagulated with 5,000 units of heparin and infusion to maintain PTT at 60–90 s. Imaging studies are ordered when they will aid or alter the plan for restoration of flow. Computed tomography (CT) angiography can be obtained in many centers at all times of the day and can

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_18, © Springer New York 2015

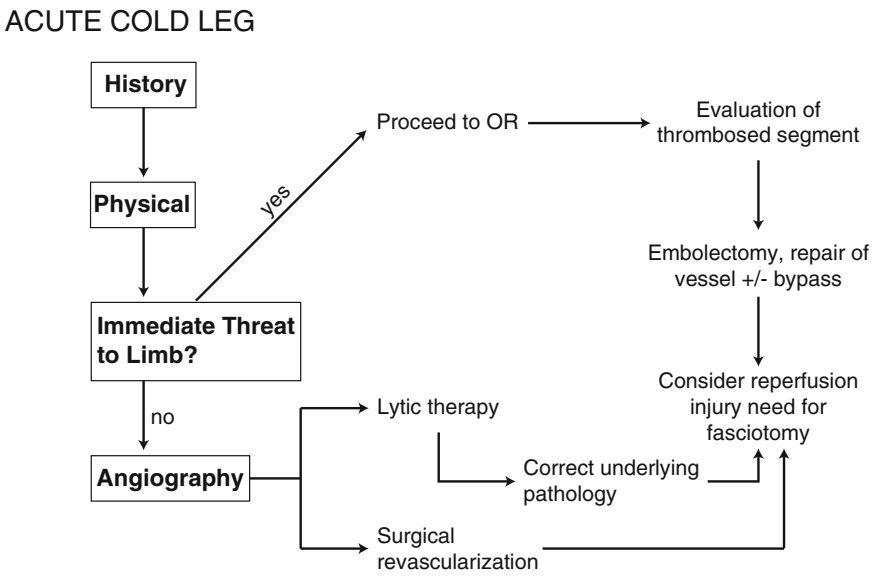

FIG. 18.1 Treatment algorithm for acute cold leg

TABLE 18.1. Classifying acute limb ischemia (Modified from Rutherford RB, Baker JD, Ernst C, Johnston KW, Porter JM, Ahn S, et al. Recommended standards for reports dealing with lower extremity ischemia: Revised version. J Vasc Surg. 1997 Sep;26 (3):517–38).

		Findings		Doppler signals	
Category of ischemia	Condition	Sensory loss	Muscle weakness	Arterial	Venous
I. Viable	Not immediately threatened	None	None	Audible	Audible
II. A. Marginally threatened	Salvageable	Minimal (toes) or none	None	Inaudible	Inaudible
B. Immediately threatened	Salvageable	Foot, calf pain	Mild to moderate	Inaudible	Inaudible
III. Irreversible	Major tissue loss, permanent nerve damage	Profound sensory loss	Paralysis	Inaudible	Inaudible

be performed rapidly, without delaying intervention. Ultrasound is noninvasive and does not require contrast but may not fully evaluate proximal arterial segments such as the aorta or iliac vessels. Ultrasound may not be available after hours and is operator dependent. Traditional arteriography provides excellent arterial imaging. Thrombolysis can be initiated at the same time, when indicated.

A patient with an immediately threatened limb is taken to the operating room. The patient is positioned, prepped, and draped such that the ischemic vascular segment can be accessed, angiography can be performed, and alternative inflow sites are accessible. For example, if the right leg is ischemic due to iliac dissection after arterial catheterization and ipsilateral iliac flow cannot be restored, preparations for a left-to-right femorofemoral bypass should have already been made.

In acute embolic disease, the embolism most frequently lodges at a point of narrowing or vessel bifurcation. Bedside duplex ultrasound may aid in rapid identification of the thrombus location. The vessel is then exposed, controlled, and explored. Balloon embolectomy via a transverse arteriotomy can be used to retrieve embolic material. On-table angiography should be used to confirm distal patency. This may reveal acute-on-chronic disease, and distal bypass may be necessary. Venous conduit is chosen when possible.

The treatment decision for the viable or marginally threatened limb is more complex. These patients often have chronic disease and developed collaterals. Angiography and surgical correction may be undertaken. Catheter-directed thrombolysis is another frequently chosen option. After angiography, a catheter is placed in the area of occlusion, and lytics are used to attempt to open the occlusion. Flow is restored, and often an underlying cause of the occlusion can be ascertained, and repaired in a more elective setting.

Reperfusion injury may occur when ischemic tissue is revascularized. The changes of injury increase with the duration of ischemia. Significant rhabdomyolysis may occur, leading to kidney injury. Hyperkalemia can also occur. Four-compartment fasciotomy must be considered after reperfusing ischemic muscle.

19
Myocardial Infarction

Li Shien Low and Clifford J. Kavinsky

Introduction

According to the American Heart Association update of the 2010 Heart Disease and Stroke Statistics, 17.6 million people in the United States have coronary artery disease (CAD). With advancements in interventional cardiology and aggressive medical management of CAD, the mortality rate has continued to decline over the past 40 years. However, CAD remains responsible for a third of all deaths in individuals over the age of 35. It is estimated that an American will have a coronary event every 25 s, with someone dying every 60 s.

Myocardial infarction (MI) is defined as a pathological event causing myocardial ischemia with evidence of myocardial injury or necrosis. The inciting event is most often due to atherosclerotic plaque disruption, which leads to the activation of the clotting cascade, causing thrombus formation and coronary artery occlusion. The majority of "culprit" lesions occlude less than 50 % of the arterial lumen prior to the event. The thin, unstable fibrous cap overlying a lipid-laden core fissures at a weak point and exposes blood in the lumen to the plaque's underlying subendocardial matrix. This leads to platelet activation, adhesion, and aggregation, as well as thrombin generation and fibrin cross-linking. Once the thrombus is formed in the artery, blood flow is impeded, and the myocardial territory supplied by that artery begins to necrose. The most crucial step in management is prompt recognition, as rapid reperfusion is essential to optimize myocardial salvage and reduce mortality – "time is muscle."

Diagnosis

The diagnosis of an acute MI is based on three critical criteria: (1) ischemic symptoms, (2) characteristic EKG changes, and (3) elevation in cardiac-specific markers (Fig. 19.1). A targeted history should elicit time of onset and nature of pain, which is typically a squeezing, crushing, or pressure-like sensation, and may radiate to the left arm or jaw. The associated symptoms include shortness of breath, nausea, vomiting, diaphoresis, palpitations, and lightheadedness. Diabetic patients, women, and the elderly are particularly prone to atypical presentations or "angina equivalent" such as isolated arm, jaw pain, epigastric discomfort, and acute mental status change.

The electrocardiographic (ECG) diagnosis of acute MI is made by demonstrating *ST-segment elevation* of greater than or equal to one millimeter ($\geq$0.1 mV) in two or more contiguous leads or the presence of a *new left bundle branch block* (LBBB) pattern in a patient experiencing characteristic acute chest symptoms. Reciprocal changes with ST depressions may be seen in appropriate leads (Table 19.1). One should also keep in mind that 50 % of the patients presenting with inferior MI (ST-segment elevation in leads II, III, and aVF) will have right ventricular involvement and should have a dedicated right-sided ECG ("reverse-mirror test"), which may demonstrate ST-segment elevation or "Q" waves in the right-sided precordial leads, particularly V3R or V4R.

Fifty percent of the ECGs in patients presenting with an acute MI do not display ST-segment elevation and are non-diagnostic. They may demonstrate only ST depression, T-wave changes, or other nonspecific features. For example, an occlusion of the left circumflex artery often presents without ST-segment elevation. Since this is the case, recent changes have been made that designate infarctions with the aforementioned typical pattern as "ST-segment-elevation MIs" and others that present with elevations in cardiac markers but without ST elevation as "non-ST-segment-elevation MIs" (Fig. 19.2). The terms Q-wave and non-Q-wave MI have been supplanted by this new terminology.

Recent advances in the understanding of the pathogenesis of MI contributed to the rapidly evolving treatment strategies of a non-ST-segment-elevation MI, and current therapy involves the use of heparin, glycoprotein IIb/IIIa inhibitors, and percutaneous intervention. Further elaboration on this

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_19, © Springer New York 2015

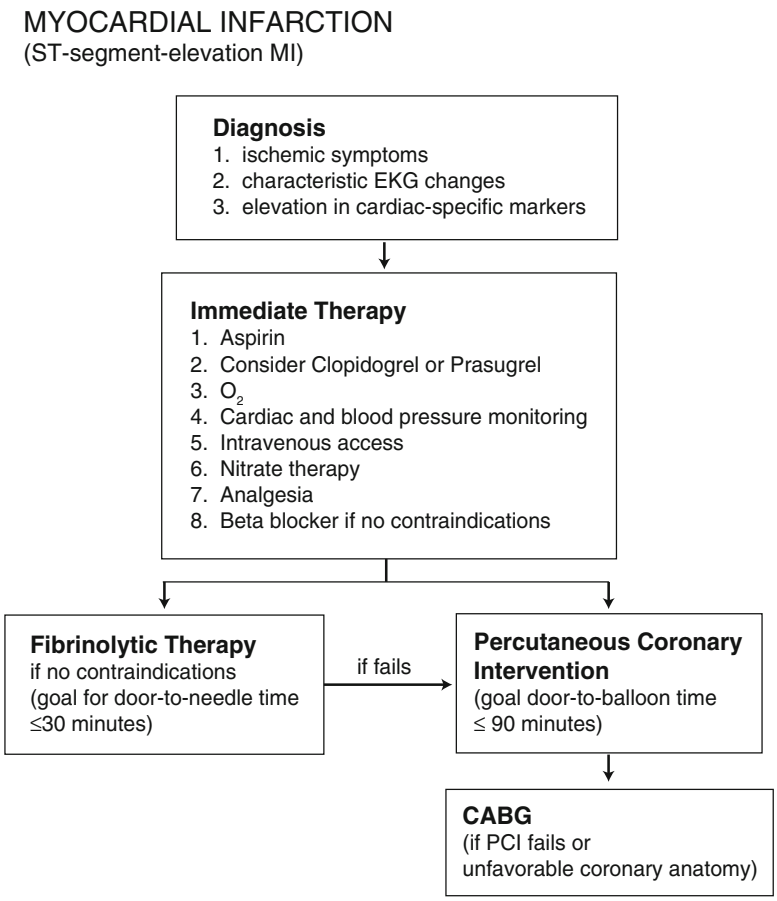

FIG. 19.1 Treatment algorithm for myocardial infarction. *EKG* electrocardiogram, O_2 oxygen, *IV* intravenous, *PCI* percutaneous coronary intervention, *CABG* coronary artery bypass grafting

TABLE 19.1. ECG leads in relation to coronary artery and myocardial territory.

Leads	Coronary artery	Myocardial territory
I, aVL	Diagonal or proximal LCx	High lateral wall
II, III, aVF	RCA or LCx	Inferior
V1–V2	Proximal LAD	Septal
V2–V4	LAD	Anterior wall
V5–V6	LCx	Lateral wall

topic is beyond the scope of this chapter, and the discussion that follows will focus on ST-segment-elevation MI.

Cardiac markers should be included in the immediate blood work. Troponin I is released in response to cardiomyocyte cell death and is extremely sensitive but takes 6–12 h from symptom onset to begin to rise. Myoglobin is a nonspecific indicator and can be elevated by any skeletal muscle or myocardial injury, but it rises within 1–2 h of symptom onset, therefore rendering it a valuable diagnostic tool for patients arriving early in the course of their chest pain. CKMB is a fractionated portion of the total creatine kinase (CK), and an elevation in the MB index (depending on a given laboratory's standards, over 2–5 % of the total CK) is diagnostic of an MI. Troponin I elevations can be prolonged in patients with renal insufficiency.

Echocardiography performed at the bedside can also be of diagnostic assistance in patients with chest pain but non-diagnostic ECGs. The presence of a new left ventricular regional wall motion abnormality on two-dimensional imaging is indicative of ongoing ischemia or infarction.

Immediate Therapy

Immediate therapy should include the following:

1. Aspirin 325 mg, chewed or crushed
2. Consider clopidogrel 600 mg load or prasugrel 60 mg load
3. Supplemental oxygen via nasal cannula
4. Cardiac and blood pressure monitoring
5. Intravenous access (with simultaneous blood draw for lab studies)
6. Nitrate therapy (sublingual nitroglycerin followed by intravenous)
7. Analgesia (morphine; or meperidine if morphine allergy present)

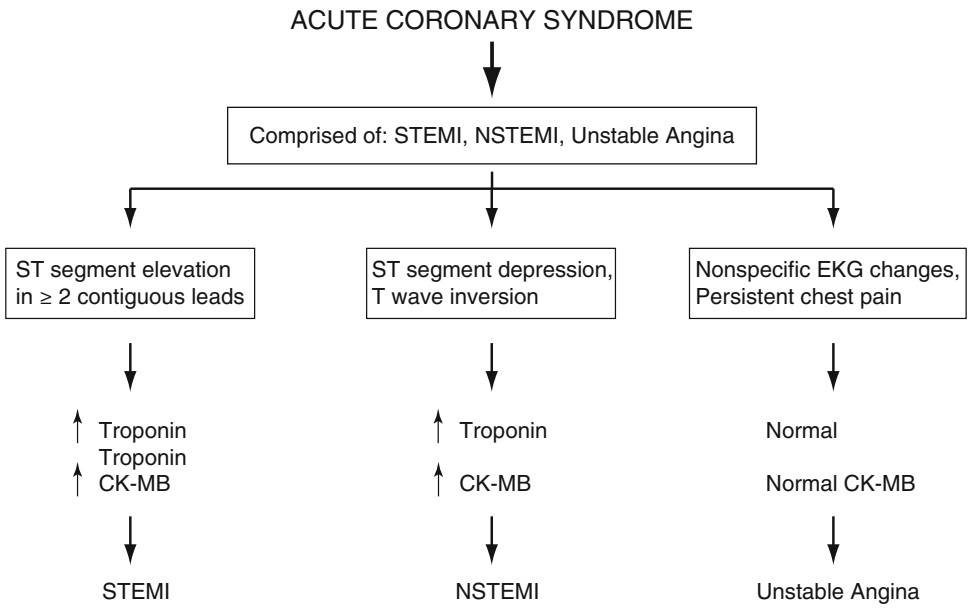

FIG. 19.2 Acute coronary syndrome. *STEMI* ST-segment-elevation MI, *NSTEMI* non-ST-segment-elevation MI

Consider beta-blocker therapy (IV metoprolol up to 15 mg in three divided doses or IV atenolol up to 10 mg in two divided doses). Contraindications include heart rate below 60 and systolic blood pressure below 100 mmHg.

Once the diagnosis of acute MI is established, reperfusion therapy is the immediate goal. The amount of myocardial territory that can be rescued from necrosis is directly related to the rapidity with which reperfusion is established. The various possibilities used to achieve reperfusion are fibrinolytics, percutaneous coronary intervention (PCI), and coronary artery bypass grafting (CABG).

Fibrinolytics

Fibrinolytics are given systematically through the IV and serve to dissolve the newly formed thrombus in the coronary artery (Table 19.2). Door-to-needle goal time is ≤30 min. There is little benefit to pharmacologic reperfusion if the patient's chest pain has been continuously present for more than 12 h.

Absolute contraindications include (1) previous hemorrhagic stroke at any time or ischemic stroke within 3 months, (2) known intracranial neoplasm, (3) active internal bleeding, and (4) suspected aortic dissection.

Relative contraindications include:

1. Uncontrolled severe hypertension (BP > 180/110 mmHg)
2. History of cerebrovascular event or other intracranial pathology not listed above
3. Anticoagulation (INR > 2–3) or known bleeding diathesis
4. Recent (2–4 weeks) trauma, including head trauma
5. Traumatic or prolonged (>10 min) CPR or major surgery (<3 week)
6. Noncompressible vascular punctures
7. Recent (2–4 weeks) internal bleeding
8. Prior allergic reaction to streptokinase
9. Pregnancy
10. Active peptic ulcer disease

TABLE 19.2. Approved fibrinolytics and doses.

Fibrinolytic	Dose
Streptokinase	1.5 MU in 30–60 min
Alteplase	100 mg in 90 min
Reteplase	10 U × 2 over 30 min

Intravenous heparin (60 U/kg bolus, followed by infusion of 12 U/kg per hour for aPTT 60–90 s) or low molecular weight heparin (1 mg/kg SC q12 h) should be administered along with alteplase and reteplase. Reperfusion failure (defined as <50 % ST-segment resolution on follow-up ECG at 60 to 90 min) is an indication for emergent rescue PCI.

PCI

PCI is often the first choice for reperfusion therapy depending on practices in the local medical community and availability of catheterization facilities and experienced operators. Goal for door-to-balloon time is ≤90 min. If this goal cannot

be met, fibrinolytic therapy is preferred. The population that benefits most from PCI as compared to fibrinolysis consists of those with anterior MI, persistent tachycardia, cardiogenic shock, or age over 70 years.

CABG is performed in the acute setting when reperfusion therapy by PCI fails or is found to be impossible after visualization of the coronary anatomy by angiography. It is reserved for such situations in the first 4–6 h after symptom onset and is not considered first-line therapy.

Prognosis

In the 1960s, in-hospital mortality rates for MI were as high as 35 %. Since that time, evolution in understanding the disease process coupled with advances in therapy has produced a steady decline from 10 to 15 % in the 1980s to the current low of 5–8 %. In general, indicators of a poorer prognosis are advanced age, cardiogenic shock, tachycardia, and location of infarction in the anterior left ventricular wall.

20
Abdominal Aortic Aneurysm

Bernadette Aulivola and Mike Malinowski

Demographics and Natural History

An aneurysm is defined as a focal enlargement of an artery to greater than 1.5 times its normal diameter. Given that the normal size of the adult aorta ranges from 1.2 to 2.4 cm, *an abdominal aortic diameter measuring larger than 3 cm is generally considered to be an abdominal aortic aneurysm (AAA)*. The prevalence of AAA is highest in white males. The aneurysm detection and management (ADAM) study demonstrated the smoking-adjusted prevalence of AAA to be 1.41 % in Caucasian versus 0.55 % in African-American men greater than 64 years of age. The pathogenesis of aneurysm formation likely involves arterial wall degeneration by atherosclerosis, chronic inflammation, and increased proteolytic enzyme activity (Fig. 20.1). Risk factors include advanced age, male sex, tobacco use, emphysema/chronic obstructive pulmonary disease (COPD), atherosclerosis, and hypertension (HTN). First-degree relatives of those diagnosed with AAA have a 15–20 % incidence of AAA and should therefore be advised to undergo ultrasound screening. *Current screening recommendations include performing abdominal aortic ultrasound in all males over 65 years of age with a history of smoking or a family history of AAA*. Approximately 80 % of AAAs are associated with smoking, which increases a patient's risk by 7.6 times in active and 3.0 times in ex-smokers when compared to nonsmoking cohorts.

Nearly 90 % of aortic aneurysms are located in the infrarenal aorta; only 5 % requiring repair are suprarenal, necessitating reimplantation of at least one renal artery. Thoracoabdominal aneurysms are rare, accounting for only 2 %. Approximately 25 % have concomitant iliac artery aneurysms and 10 % have popliteal artery aneurysms. Conversely, 40 % of patients diagnosed with popliteal artery aneurysms are found to have an AAA, making screening for these patients essential.

The most dreaded complication of AAA is rupture, the risk of which is best predicted by the maximal cross-sectional diameter of the aneurysm (Table 20.1). Patients with COPD and HTN have a higher aneurysm rupture risk, conceivably due to the intra-abdominal and intravascular changes in pressure dynamics associated with these comorbidities. If left alone, they tend to enlarge and may eventually rupture. *Up to 80 % of patients with ruptured AAA will not survive the event*. Only 50 % of patients with a ruptured AAA will even make it to the hospital alive and up to 50 % of these patients will die during the course of their hospitalization.

Approximately 80 % of all AAAs are discovered incidentally on abdominal ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), or plain radiograph. Physical examination can detect only 50 % of AAAs measuring 4–5 cm and 75 % of those measuring >5 cm in diameter. Obese body habitus may make palpation of the abdominal aorta challenging to perform. Most AAAs are asymptomatic until they rupture. Some patients present with abdominal or back pain in the absence of evidence of aneurysm rupture on CT scan. These patients may complain of tenderness to palpation of the aneurysm as well. Assuming all other causes of pain are ruled out, urgent AAA repair should be considered. Rarely, AAAs can present with symptoms related to distal embolization of intraluminal thrombus. These symptoms occur in less than 5 % of patients with AAA and can manifest as what is known as "blue toe syndrome." In such cases, thrombus or atherosclerotic plaque embolize to the arteries of the distal extremity resulting in focal areas of ischemia that may progress to gangrene. Some such cases can be associated with aneurysms smaller than 5 cm and may be an indication for repair of a small aneurysm.

Imaging and Surveillance

Ultrasound Versus CTA Versus DSA

Abdominal ultrasound remains the simplest and most cost-effective modality for screening and initial diagnosis. CT and MRI allow for post-processing three-dimensional

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_20, © Springer New York 2015

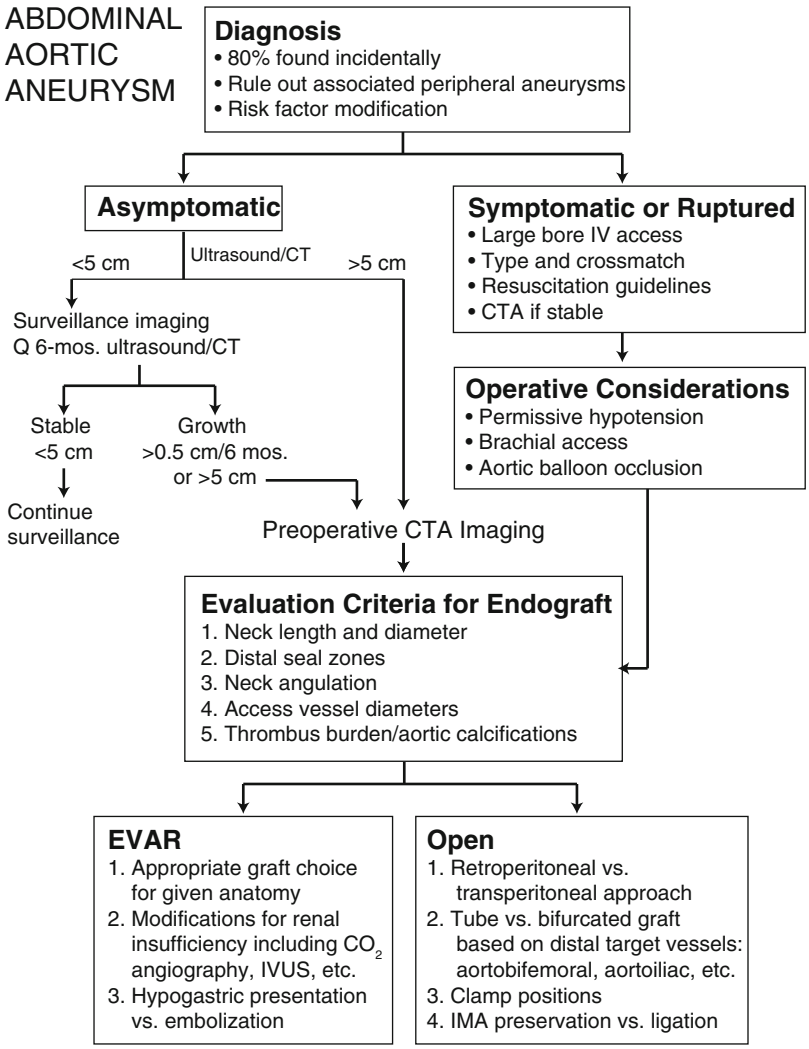

FIG. 20.1 Algorithm for treatment of abdominal aortic aneurysm (AAA). *CT* computed tomography, *IV* intravenous, *CTA* computed tomography angiography, *EVAR* endovascular aneurysm repair, *IMA* inferior mesenteric artery

TABLE 20.1. Risk of AAA rupture.

Cross-sectional AAA diameter (cm)	Rupture risk (% per year)
<4	<1
4–5	1–3
5–7	6–10
>7	>20

reconstructions to assist in operative planning. Patients with a small AAA that is adequately imaged on ultrasound should undergo surveillance ultrasound every 6 months or annually; once an AAA nears the size that would necessitate repair, patients should undergo IV contrast-enhanced spiral CT angiography (CTA) with 2–3 mm cuts to obtain accurate information regarding size and extent. MRI may be used if a contraindication to administering intravenous iodinated contrast dye exists; however, fine-cut CTA remains the imaging modality of choice. Standard digital subtraction angiography (DSA) has specific limitations; it does not accurately demonstrate maximal aneurysm diameter given that many aneurysms contain intraluminal thrombus. Angiography does, however, provide accurate information regarding renal artery number, location, and orientation; mesenteric artery patency; internal iliac artery location and patency; and presence of aortoiliac occlusive disease. Angiography should be considered in patients with juxtarenal or suprarenal AAA, presumed renovascular HTN, chronic mesenteric ischemia, or lower extremity ischemia. In many cases, however, CTA can provide as much information as DSA and it has the advantage of being less invasive. CTA is an essential tool in evaluating the patient with an AAA for open or endovascular aneurysm repair (EVAR).

Indications for Repair

Decisions on elective aneurysm repair should take into account comorbid medical conditions and the risk of aneurysm rupture. *AAAs with diameter growth rates of greater than 0.5 cm in 6 months or 1.0 cm in one year should be*

considered for repair. AAAs *larger than 5–5.5 cm warrant elective repair*, as the rupture risk outweighs the operative risk in patients who are reasonable operative candidates. Because of the high incidence of rupture of large AAAs (>6 cm), even patients with elevated cardiac risk should be considered for repair. Patients with abdominal or back pain without other obvious causes in the presence of AAA should undergo urgent repair. This includes patients without evidence of rupture on CT scan, given the possibility of impending rupture. AAA rupture is a definite indication for emergent repair, without which, the mortality rate is 100 %.

Preoperative Considerations

Coronary artery disease (CAD) is the most accurate predictor of perioperative morbidity and long-term mortality, and it is present in almost two-thirds of AAA patients. Since a large proportion of perioperative and late deaths are due to CAD, preoperative assessment of cardiac status is essential. Stress echocardiography or adenosine thallium scans should be considered, particularly in patients with a history of myocardial infarction (MI), CAD, ventricular arrhythmia, diabetes, or angina. Stress testing should be followed by cardiac catheterization to further investigate the severity of CAD in those with reversible ischemic defects. When indicated, coronary artery bypass grafting or percutaneous coronary intervention prior to AAA resection may lead to improved survival.

AAA Repair

Ruptured AAA Repair

In patients with hemodynamic instability, emergent repair should be performed. Endovascular repair may be considered if stent graft devices are readily available. To minimize blood loss, the patient may be resuscitated with permissive hypotension from the time of the rupture diagnosis until the time of aortic control. Systolic blood pressure in the 80 mmHg range is acceptable. Crystalloid infusion should precede blood transfusion if possible. Packed red blood cells, platelets, and fresh frozen plasma should be available. The patient should undergo arterial and central venous line placement and should be prepped and draped with the surgeon poised to operate. Induction of general anesthesia may precipitate hemodynamic collapse. Proximal aortic control should be a priority, regardless of whether repair of the ruptured AAA is open or endovascular. Options for proximal control include supraceliac clamping and aortic balloon occlusion via a brachial or femoral access site. Aortic balloon occlusion requires a 12 French sheath; therefore, when brachial access is used, a cutdown is generally preferred. Aortic control may allow resuscitation efforts to be continued by the anesthesiologist.

Elective AAA Repair

Patients undergoing AAA repair should receive preoperative antibiotics to reduce the risk of graft infection. Epidural catheter placement for postoperative pain management should be considered. Preoperative bowel preparation is generally not necessary. For open operative intervention, the optimal aortic approach is through either a midline laparotomy with transperitoneal dissection or a left retroperitoneal exposure. The latter approach provides improved pararenal exposure and is associated with decreased rates of postoperative respiratory complications and ileus; however, exposure of the right iliac artery is more difficult. Alternatively, although rare, a right retroperitoneal approach may be used in cases where a stoma would interfere with this approach from the left. When open repair is chosen, relative indications for the retroperitoneal approach are a history of multiple previous transperitoneal surgeries, an abdominal stoma, a horseshoe kidney, an inflammatory AAA, or an expected need for renal revascularization.

Proximal and distal exposure is performed with minimal manipulation of the aneurysm to reduce the risk of distal embolization. For infrarenal AAAs, proximal control is usually obtained just distal to the renal arteries, which is usually marked anteriorly by the crossing left renal vein. In cases of more proximal aneurysm extension – such as in juxtarenal or suprarenal aneurysms – proximal clamp position may be placed in the suprarenal or supraceliac positions. For suprarenal clamping, division of the left renal vein can be performed with care taken to preserve gonadal vein collaterals. Supraceliac clamp placement requires division of the gastrohepatic ligament and right crus of the diaphragm. Heparin (80 units/kg) is administered intravenously prior to vascular clamping; most surgeons do not administer heparin in cases of ruptured AAA. In order to minimize the risk of embolization, distal vascular clamps are usually applied prior to proximal aortic clamp placement. The aneurysm sac is then opened, thrombus is removed, and lumbar arteries are suture ligated from within the sac to control backbleeding. Autotransfusion devices are helpful in minimizing banked-blood transfusion intraoperatively. The inferior mesenteric artery (IMA) should be evaluated for the need for reimplantation onto the aortic interposition graft but can usually be ligated at its origin. *Relative indications for reimplantation of the IMA include factors that restrict pelvic collateral flow such as internal iliac artery occlusion or prior colon resection*. Typically, a tube graft is placed, but a bifurcated graft may be required if iliac artery aneurysms or external iliac

artery occlusive disease are present. When extensive external iliac occlusive disease is present, distal graft limbs may be tunneled to the groin for femoral anastomosis. Once the graft is sutured in place, flow is restored first to the internal then to the external iliac arteries to divert potential emboli from the lower extremities. The aneurysm sac is closed over the graft to prevent aortoenteric fistula formation. Visual inspection of the left colon should be performed to evaluate for ischemia. Distal flow is assessed by palpating femoral and distal pulses.

Postoperative care includes cardiac afterload reduction to reduce the risk of bleeding and myocardial ischemia. Beta-blockade should be considered to reduce postoperative cardiac morbidity. Potential complications include myocardial infarction (3–5 %), stroke, renal failure, lower extremity ischemia, colon ischemia (2–6 %), pulmonary insufficiency, infection, and spinal cord ischemia. Colon ischemia usually presents in the first 3 days, and may manifest as blood-tinged stools, left lower quadrant pain, fever, leukocytosis, and tachycardia. These findings should trigger evaluation with flexible sigmoidoscopy followed by intravenous antibiotics and possibly exploratory laparotomy if transmural ischemia is suspected or if signs of peritonitis are noted. Perioperative mortality with elective open AAA repair is 5 % of which two-thirds is attributable to cardiac complications.

Endovascular AAA Repair (EVAR)

All patients with an AAA diagnosis should be considered for endovascular repair. One relative contraindication to EVAR is renal insufficiency, given the use of intravascular contrast agents for imaging. Several alternatives to standard contrast agent use have made EVAR more feasible in patients with preexisting renal insufficiency. These include use of intravascular ultrasound, carbon dioxide angiography, and use of postoperative noncontrast CT in combination with duplex imaging to assess for AAA sac diameter and endoleak presence.

Anatomic factors play a critical role in determining if a patient is a suitable candidate for EVAR. These anatomic criteria relate to access (ability to pass the graft into position) and aortoiliac anatomy (ability to obtain seal zones of the proximal and distal aspects of the graft). Among the most critical of these criteria are:

1. Infrarenal aortic neck diameter (19–32 mm)
2. Infrarenal aortic neck length (>10 mm)
3. Infrarenal aortic neck angulation (<60°)
4. Femoral and iliac lumen diameter (>7–8 mm for main body deployment)

TABLE 20.2. Favorable vascular anatomy for endovascular stenting.

1. A normal infrarenal segment of aorta at least 10 mm in length and no more than 32 mm in diameter
2. Minimal iliac artery inner diameter of 7 mm
3. Angulation of proximal aortic neck of less than 60°
4. Angulation of iliac arteries of less than 90°

EVAR may be performed with open femoral exposure or percutaneous techniques. Favorable vascular anatomy for endovascular repair is summarized in Table 20.2. The long-term benefits of endovascular repair in patients considered high risk for open AAA repair are questionable. Reduction in perioperative morbidity and shortened hospital stay have helped this AAA repair technique gain popularity. Complications of EVAR include groin wound complications, contrast-induced nephropathy, and endoleaks. Endoleaks are the presence of persistent blood flow within the aneurysm sac. Four main types of endoleaks have been described:

- Type I leaks consist of flow into the aneurysm sac via proximal (IA) or distal (IB) seal zones of the endograft.
- Type II endoleaks consist of retrograde flow from aortic branch vessels including lumbar, internal iliac, accessory renal, middle sacral, and inferior mesenteric arteries.
- Type III endoleaks originate from junctions between graft components or tears in the graft wall.
- Type IV endoleaks originate from graft wall fabric porosity or suture holes and have become less common with newer graft technology.

In addition to these four types, an entity referred to as "endotension" consists of high AAA sac pressure without a demonstrable evidence of persistent flow into the AAA sac. Approximately 70 % of type II endoleaks resolve spontaneously within a month of graft placement. Type II leaks are thought to be the most benign as they are not thought to provide arterial pressure to the AAA sac. When a type II endoleak is accompanied with aneurysm sac expansion on surveillance, repair is indicated, however. EVAR repair requires lifelong follow-up with CT scanning, plain radiographs, and/or ultrasound imaging. *Approximately 30 % of EVARs will require a secondary procedure at some point in time.*

Inflammatory Aneurysms

Inflammatory aneurysms account for 5–10 % of all AAAs and have distinct characteristics that differ from typical atherosclerotic AAAs. They tend to occur in younger patients and are associated with an increased erythrocyte sedimentation rate.

CT scan imaging demonstrates distinct findings, including a thickened aortic wall with contrast enhancement. The inflammatory AAA is usually associated with retroperitoneal fibrosis and adherence of the aorta to surrounding structures such as the duodenum. Risk factors include smoking and male sex. Although the exact etiology of inflammatory aneurysms remains unclear, infection is not thought to play a role. Repair criteria are the same as in any AAA. Repair should be performed with EVAR if possible, and when open repair is indicated, a retroperitoneal approach is recommended, as it avoids risk of injury to adherent structures to the aneurysm.

Postoperative Surveillance

After open AAA repair, the patient has minimal risk of progressive aneurysm formation of the aorta proximal to or distal to the repair. Aortoduodenal fistula complicates less than 1 % of cases. Surveillance CT scan every 5 years should be considered. EVAR cases should undergo surveillance imaging with CTA at 1, 3 or 6, 12, 18, and 24 months, then annually thereafter, assuming stable. Evaluation with plain radiograph, noncontrast CT, and duplex may be considered as alternatives to contrast-enhanced CT.

21
Dialysis Access

Oyedolamu K. Olaitan and Edward F. Hollinger

Introduction

Although the incidence has largely plateaued, the prevalence of end-stage renal disease (ESRD) in the United States continues to increase. As of December 31, 2011, there were approximately 388,000 patients on hemodialysis (HD), 31,200 patients on peritoneal dialysis (PD), and 185,000 patients with a functioning kidney transplant. Renal transplantation is the preferred treatment for ESRD; however, the number of potential recipients continues to outstrip the number of available kidneys. As of 2013, there were about 98,000 patients on the waiting list for kidney transplant, while only between 16,000–17,000 transplants are performed each year (Organ Procurement and Transplant Network Data, November 2013). The disparity between donors and recipients leads to increasing waiting times for kidney transplant (averaging 5–7 years in some areas) with a concomitant increasing need for renal replacement therapy, both for those waiting for kidney transplant and for those patients who are not candidates. The cost of ESRD is significant, with nearly $50 billion in direct costs in 2011.

Indications for Dialysis

Urgent dialysis can be used to treat sequelae of acute kidney injury including fluid overload, hyperkalemia or acidosis, refractory hypertension, pericarditis, or mental status changes (Fig. 21.1). If the kidneys are expected to recover, a temporary central venous catheter can be inserted either in the internal jugular (IJ) vein or the common femoral vein. The right internal jugular vein is preferred because of the more linear pathway when directing the catheter tip to the cavoatrial junction. Placement of dialysis (or other) catheters in the subclavian veins should be avoided as it may lead to venous sclerosis and occlusion, which can preclude subsequent placement of arteriovenous (AV) fistulas or grafts in the ipsilateral arm.

Chronic renal replacement therapy (RRT) generally is initiated when the patient reaches a glomerular filtration rate (GFR) of 10–15 mL/min/1.73 m^2, when they have symptomatic fluid overload, electrolyte or acid-base imbalance, or significant clinical sequelae such as neurologic changes, uremic pericarditis, or chronic gastrointestinal (GI) symptoms. Of patients beginning renal replacement therapy, more than 90 % of patients beginning chronic RRT begin with hemodialysis (HD). The rest begin peritoneal dialysis (6.5 %) or receive a kidney transplant preemptively (2.5 %). Despite increased emphasis on early diagnosis and referral for nephrology care, in 2011 more than 40 % of patients diagnosed with ESRD had not seen a nephrologist until beginning renal replacement therapy. Ideally, patients should be referred for access placement well in advance of the time of initiation of dialysis to give time for the fistula or graft to mature or to allow the peritoneum to seal around the peritoneal dialysis catheter insertion site.

Because of improved outcomes and decreased costs, there is increasing emphasis on preemptive placement of long-term dialysis access (arteriovenous fistula or graft) as compared to catheter-based dialysis. Of patients without prior access to nephrology care, over half began HD with a catheter, as compared to less than one-third of patients with at least one year of pre-dialysis nephrology follow-up. Many of these patients will subsequently transition to AV fistulas or grafts (for hemodialysis) or peritoneal dialysis. Therefore, consideration of long-term dialysis access is important when placing a temporary (non-cuffed) or tunneled "permanent" cuffed dialysis catheter.

Access Planning

Patients with stage 4 CKD (GFR 15–29 mL/min/1.73 m^2) and those that are expected to require dialysis within 6 months should be referred for access planning. The decision

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_21, © Springer New York 2015

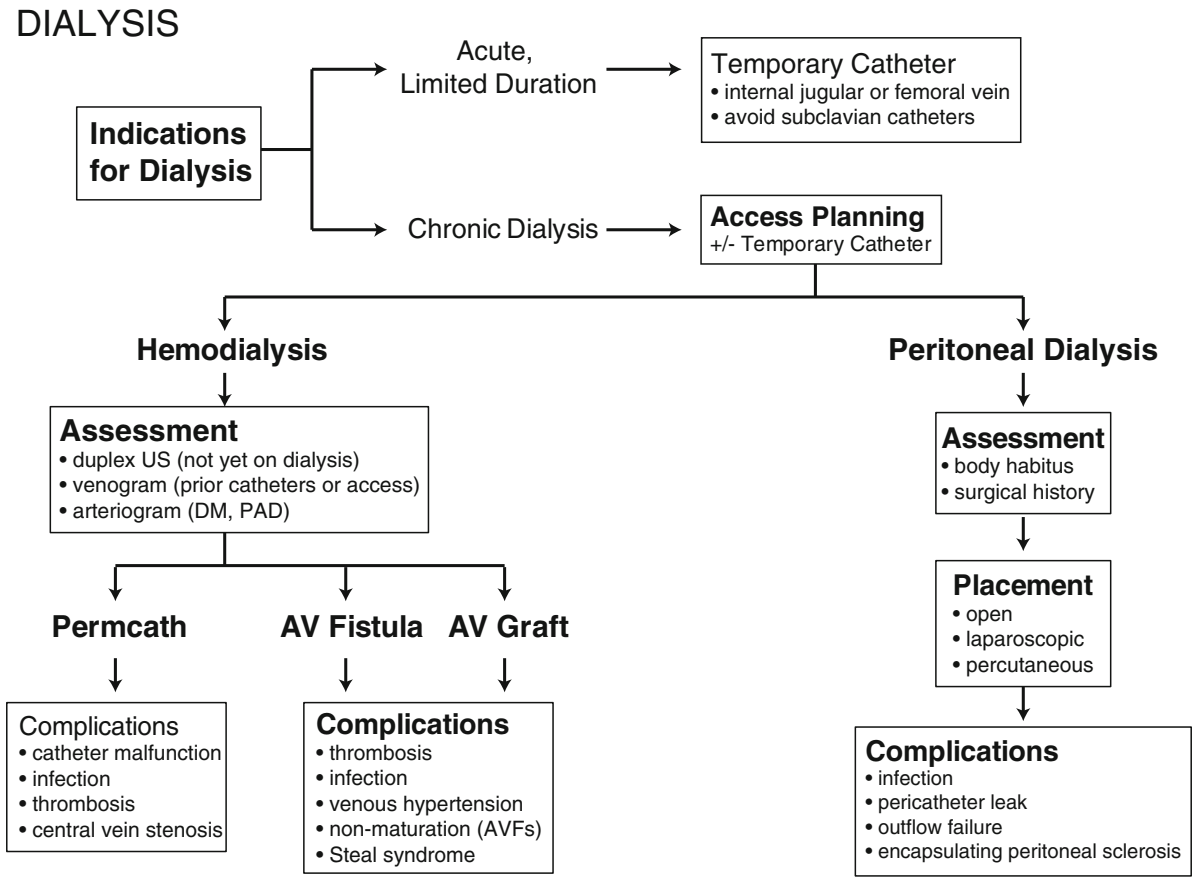

Fig. 21.1 Algorithm for initiating dialysis. *US* ultrasound, *DM* diabetes mellitus, *PAD* peripheral artery disease, *AV* arteriovenous, *AVF* arteriovenous fistula

whether to pursue hemodialysis or peritoneal dialysis should be made with input from the nephrologist and surgeon and based on the patient's preference and the medical and surgical feasibility. The patient also should be referred for evaluation for renal transplantation (preferable when GFR is approaching 20 mL/min/1.73 m^2, which is when they can start accumulating time on the transplant waiting list).

Hemodialysis Access

HD can be performed via a catheter, arteriovenous fistula (AVF), or an arteriovenous graft (AVG). For adequate dialysis, the access must support blood flow rates in excess of 300 mL/min. A fistula is created by anastomosing a native vein to an artery. Over the next several weeks to months, the increased pressure and blood flow in the vein leads to increased diameter and wall thickness, so-called maturation. An AV graft connects the artery and vein by interposition of a synthetic (usually expanded polytetrafluoroethylene [PTFE]) conduit. The National Kidney Foundation's Kidney Disease Outcome Quality Initiative (NKF/KDOQI) recommends AVF use of greater than 50 % in new patients and greater than 40 % in patients already on hemodialysis. This initiative is supported by the Centers of Medicare and Medicaid Services (CMS) through the National Vascular Access Improvement Initiative (Fistula First), because AVFs have higher patency rates and lower incidence of infection, interventions, hospitalization, and death when compared with other types of access.

Preoperative Assessment

Successful placement of an AV fistula or graft requires arterial inflow and venous outflow. For an AVF, an appropriate quality vein is also required. The selected vein must be amenable to percutaneous access, either in its anatomic location or after surgical superficialization or transposition. The patient's medical history should be carefully evaluated to assess fitness for surgery as well as risk factors for central venous occlusion, including:

- Prior central venous catheters; especially subclavian catheters
- Prior central venous stenosis or thrombus

- Unilateral upper-extremity swelling
- Prior fistula placement and cause of failure

The fistula or graft is preferentially placed in the patient's nondominant arm, so that the patient can have the dominant hand free during dialysis.

Apart from the general examination to assess fitness for surgery/anesthesia, emphasis should be placed on examination of the venous and arterial systems. Arterial examination includes assessing skin integrity and perfusion, capillary refill, quality of pulses, and blood pressure and performing an Allen test to ascertain a complete palmar arch. The arm should be examined for any obvious veins; this can be assisted by using a light venous tourniquet above the examined vein.

The size and patency of the veins can be assessed with Doppler ultrasonography (US) or contrast venography. Venous ultrasound provides a good representation of the size, location, and quality of the extremity veins and arteries. It is generally not very useful in assessing the central veins (subclavian, innominate, SVC). Contrast venography is less operator dependent and delineates the central vasculature (and any potential outflow stenosis) better than venous duplex. Carbon dioxide can be used as a contrast agent in patients not yet on dialysis to avoid nephrotoxicity. If the physical exam is inconclusive or if risk factors are present, arterial duplex can be used to evaluate the arterial inflow as well as digital perfusion. This is especially important if an upper arm or brachial artery inflow is planned as they are associated with a higher incidence of distal ischemia. When the distal radial artery is to be used for inflow, the palmar arch should be assessed, either by physical exam (Allen test) or with duplex ultrasound. A careful assessment for arterial insufficiency is mandatory prior to placing any lower-extremity access because of the risk for limb ischemia.

Permanent Catheters

The use of a large-bore tunneled "permcath" in the IJ or femoral vein allows dialysis immediately following catheter placement. Placement with imaging guidance (US/fluoroscopy) is highly recommended, especially in patients with multiple prior catheters who may have central venous stenosis. When compared with an AV fistula or graft, catheters provide lower flow rates, poorer clearance, and more recirculation. When compared with AV fistulas, catheters are associated with a twofold increase in relative rate of death, a nearly sevenfold increase in bacteremic episodes, and higher cost and number of required secondary interventions. Complications of permcaths include catheter malfunction, infection, and central vein stenosis/thrombosis. The latter is particularly damaging as it can lead to loss of the ipsilateral extremity for future HD access.

Autogenous Primary Fistula

Initial evaluation and placement of the AVF should be 3–6 months prior to initiation of dialysis so as to allow time for maturation. A longer lead time may be required if the arterialized vein needs to be superficialized (moved closer to the surface along its anatomic route) or transposed (moved to a more accessible, typically nonanatomic location). Although many veins can be arterialized, some of the more common AVF configurations are listed below. The arterial anastomosis can be done end-(vein)-to-side (artery) or side-to-side. Typically, the later has a slightly higher incidence of steal syndrome:

- *Radiocephalic AVF* – created by connecting the radial artery and the cephalic vein. The anastomosis can be at the level of the anatomic snuff box, at the wrist or at the antecubital fossa. The radiocephalic AVF at the wrist (Brescia-Cimino fistula) is an excellent site because of its high patency rate, longevity, and low incidence of complications.
- *Brachiocephalic AVF* – anastomosis between the distal brachial (or proximal radial or ulnar) artery and the cephalic vein, usually at the antecubital fossa. This AVF also has good long-term patency; however, the cephalic vein may need to be superficialized, especially in obese patients in whom it may be too deep to cannulate.
- *Transposed basilic or brachio-brachial AVF* – can be performed as a single- or two-stage procedure. The first stage involves connection of the distal brachial or proximal radial or ulnar artery to the basilic or the brachial vein just above or (preferably) below the elbow. After adequate maturation (typically when the vein is at least 5 mm in diameter), the vein is transposed to overly the biceps muscle. This fistula also can be created as a single-stage procedure especially if the vein is of good caliber.
- *Forearm basilic vein AVF* – can be transposed as a loop or straight AVF to the proximal or distal radial artery, respectively. This option may be limited by the caliber and length of the forearm basilic vein.

Autogenous Secondary Fistulas

The basilic vein or great saphenous vein (GSV) are harvested and used as grafts in secondary sites in the same patient. Their use is infrequent because the long-term patency rate has been disappointing.

Non-autogenous Grafts

These are used when fistula options have been exhausted, when suitable veins are not available, or in obese patients (especially in the upper arm) where suitable length of

autogenous vein is not available to facilitate superficialization or transposition. The most common graft material is polytetrafluoroethylene (PTFE, Gore-Tex®), although multiple materials and configurations have been developed. Grafts should be planned using the same principles as AV fistulas, namely, they should be formed as distal as possible in the upper extremity. Tapered grafts are preferred to limit the arterial inflow and reduce the risk of arterial steal syndrome. Common insertion sites include:

- *Loop forearm graft* – one of the most common form of prosthetic access; it is constructed between the brachial artery or one of its major branches and a vein in the ACF.
- *Straight forearm graft* – usually constructed between the radial artery at the wrist and one of the antecubital veins at the elbow. The patient must have a strong radial flow and a complete palmar arch to ensure adequate graft and digit perfusion.
- *Upper arm arteriovenous grafts (AVG)* – can be used as secondary sites when forearm sites are exhausted or the more-distal arteries are too small. Can be constructed in a straight or looped configuration; usually uses the brachial artery as the inflow. Venous outflow can be to the cephalic, basilic, brachial, or axillary veins.
- *Lower-extremity AVG* – this site is less desirable because of the higher incidence of infection or arterial insufficiency compared with the upper extremity. The most common configuration is looped with inflow from the common femoral artery and outflow via the common femoral vein.
- *Hemodialysis reliable outflow (HeRO) graft* – this is a recent innovation developed to address the problem of central venous occlusion. It consists of an upper-extremity graft connected to a thoracic component (placed like an HD catheter) that extends to the cavoatrial junction. The thoracic component is placed under fluoroscopy, usually after recannulation of central venous stenosis or occlusion.

Complications of Fistulae and Grafts

- *Thrombosis* – most commonly due to outflow stenosis, often as a result of intimal hyperplasia in the outflow vein. AV grafts are more prone to stenosis, although also more amenable to percutaneous or surgical thrombectomy.
- *Infection* – nearly five times more common with PTFE graft than for autogenous vein and accounts for more than one-third of graft losses. Can present as local abscess, bacteremia, or septic emboli. Depending on the extent of infection, can be treated with partial resection (sometimes with immediate reconstitution with a jump graft around the infected area) or complete excision of the infected graft and repair of the arteriotomy.
- *Venous hypertension* – most commonly results from central venous stenosis and may lead to graft loss. Often amenable to percutaneous angioplasty or stent placement.
- *Non-maturation* – AV fistulas can fail to mature (or thrombose) due to poor arterial inflow, venous outflow, or poor quality of the arterialized vein. Most failing AVFs can be identified by physical exam at 4 weeks, and the rate of salvage can be significantly increased if intervention is initiated before the AVF clots. A contrast fistulogram (including evaluation of the arterial inflow as well as the venous outflow) can provide diagnostic information as well as offer therapeutic options such as balloon angioplasty.
- *Steal syndrome* – symptomatic extremity ischemia resulting from diversion of blood flow into the AVF or AVG. The patient may initially present with cool, pale, numb, or painful digits that can progress to nerve damage and distal necrosis. Therapy ranges from narrowing (banding) the access conduit to revascularization procedures such as distal revascularization/interval ligation (DRIL) to ligation of the access.

Peritoneal Dialysis

The prevalence of peritoneal dialysis (PD) in the United States has declined, but it remains popular with selected patients, especially pediatric ESRD patients where it is the modality of choice. The PD catheter has two cuffs, one that is typically placed just superficial to the peritoneal entry point and the other placed in the subcutaneous tunnel.

Preoperative Assessment

The PD catheter should be inserted at least 3–6 weeks before initiation of dialysis to allow adequate sealing of the catheter entry site. Because PD is performed at home, it requires a higher level of patient involvement and a robust support network. Relative contraindications to PD include prior abdominal surgery, ventral hernias, chronic constipation, obesity and inability to obtain sufficient clearance or fluid removal.

Catheter Insertion Techniques

- *Open surgical insertion* – can be performed under local anesthesia in thin patients using a small infraumbilical incision with the catheter entering the peritoneum in the midrectus. Open insertion has the lowest leak rate as the peritoneum can be sutured in a purse-string fashion around the catheter.
- *Laparoscopic insertion* – has the advantage of being able to visualize the peritoneal cavity and take down adhesions in patients who may have had previous abdominal surgery. It may have higher leak rate than the traditional open technique; however, the overall results are comparable in experienced hands.

- *Percutaneous insertion* – wherein the catheter is placed using a Seldinger technique with imaging guidance. This technique is unsuitable for obese patients and those that may have adhesions. It is associated with increased early leaks and there is potential for serious complications like intra-abdominal viscus perforation and hemorrhage.

Complications and Management

- *Infection* – can be localized to the exit site or generalized peritonitis. Patients with tunnel or exit site infections may present with local erythema, tenderness, and purulent discharge and can often be successfully treated with antibiotics. Peritonitis usually presents with generalized abdominal pain, often with systemic symptoms such as fever. The catheter should be aspirated and fluid sent for bacterial and fungal cultures. Empiric antibiotics (systemic or intraperitoneal) are started while waiting for the results of cultures. As a general rule, removal of the catheter is indicated in cases of resistant organisms or fungal peritonitis; the presence of multiple organisms should raise concern for bowel perforation.
- *Pericatheter leak* – presents as dialysis fluid leaking out the exit site and it indicates inadequate sealing of the exit site of the catheter from the peritoneal cavity. It occurs if enough time is not allowed between catheter placement and initiation of PD. A few weeks' hiatus in PD usually resolves the leak; major or persistent leak may require surgical revision.
- *Outflow failure* – retention of instilled dialysate can result from catheter malposition, intraluminal catheter occlusion (often from thrombus or fibrin), or extraluminal catheter obstruction (usually from omentum or adhesions). Malpositioned catheters often can be successfully repositioned using laparoscopy (with the initial pneumoperitoneum obtained via the PD catheter). Other extraluminal obstructions can be addressed with lysis of adhesions or omentopexy or omentectomy. However, eventual loss of peritoneal domain may restrict the long-term use of PD in patients with recurrent adhesion formation or (especially) with recurrent peritonitis.
- *Encapsulating peritoneal sclerosis* – EPS can develop during PD or several weeks or months after catheter removal. The patient typically presents with fever, increased C-reactive protein, and ileus symptoms usually followed by development of ascites. It can progress to peritoneal sclerosis and encapsulation with dense adhesions leading to bowel obstruction. Treatment includes cessation of PD (if ongoing) and steroid pulse. Refractory patients may require TPN. Many patients require enterolysis after the inflammation has subsided.

Pediatric Considerations

Peritoneal dialysis is preferred in the pediatric age group, especially if the patient has strong caregiver support. In ESRD patients weighing less than 10 kg, PD is preferred because it is technically easier to perform than HD, does not require a vascular access (technically challenging in small patients) or venipuncture, and reduces the need for compliance with strict fluid restrictions.

Part IV
Endocrine Disorders

22
Thyroid Nodules and Malignancy

Vikram D. Krishnamurthy and Katherine Heiden

Overview

The prevalence of palpable thyroid nodules in the US population is estimated to be 4–7 % in adults. By contrast, autopsy studies have shown that thyroid nodules are present in a much higher percentage of people – up to 50 % of adults – demonstrating that most nodules are rarely clinically detected. An aging population and the use of high-resolution ultrasonography have raised our awareness of non-palpable nodules. The challenge, thus, arises in differentiating nodules that are clinically significant from those that are not. Thyroid cancer remains an infrequent cause of malignancy, accounting for an estimated 1 % of cancer diagnoses per year. Although thyroid carcinoma typically follows an indolent course, some tumors are aggressive and capable of progressing to metastatic disease. This highlights the importance of early identification and appropriate treatment.

Evaluation

Evaluation starts with a thorough history and physical followed by laboratory values and imaging (Fig. 22.1). A key feature in evaluating the malignant potential of a nodule is recognizing high-risk patient groups. Risk factors for thyroid cancer include extremes of age (young or old), previous exposure to ionizing radiation, or a personal or family history of thyroid cancer or other endocrine tumors. Other important components of patient history include symptoms of hypo/hyperthyroidism, previous history of neck masses, underlying thyroid disease, or changes in phonation. The physical exam should include a methodical head and neck exam focusing on the thyroid gland and inspecting for enlargement, nodules, and associated cervical lymphadenopathy. If there is evidence of locally advanced disease (hoarseness, dysphagia), a fiber-optic laryngoscopy/bronchoscopy may be performed preoperatively in the office.

Laboratory and Imaging

The patient's thyroid hormonal status should be assessed with a thyroid function panel including TSH, free T3, and free T4 levels. Nodules can be further characterized by surgeon-performed ultrasonography (US) that provides detailed information about the size, number, location, and characteristics (i.e., solid versus cystic, vascularity) of nodules, as well as the presence or absence of abnormal cervical lymph nodes. Nodules with microcalcifications, irregular borders, or hypervascularity raise the suspicion for thyroid cancer. Lymph nodes that are calcified, hypervascular, spherical, or that have lost their hilar echogenicity are suggestive of metastatic disease. Fine-needle aspiration (FNA) of nodules can be performed under US guidance to provide pathologic diagnosis, which will be discussed in further detail in a subsequent section. In general, FNA should be performed on all nodules $\geq$1 cm.

Radioactive iodine uptake scan (thyroid scintigraphy) is an imaging study that determines functionality of nodules. Lesions with increased iodine uptake are considered "hot" and are hyperfunctioning. These very rarely pose a risk of malignancy. In contrast, lesions with decreased radioisotope concentration are considered "cold" and carry a higher risk of malignancy.

Investigation Based on Functional Status

Once a nodule has been discovered, the patient's thyroid hormonal status determines further workup and treatment. Hyperthyroid patients should be investigated with a radioactive iodine uptake scan. If the nodule is "hot," it can be ablated by one of three methods – radioactive iodine ablation, antithyroid medications, or surgical resection – and it does not warrant FNA. If the nodule is "cold" (in the background of homogeneous increased iodine uptake), the patient should undergo FNA of the nodule to determine necessary treatment.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_22, © Springer New York 2015

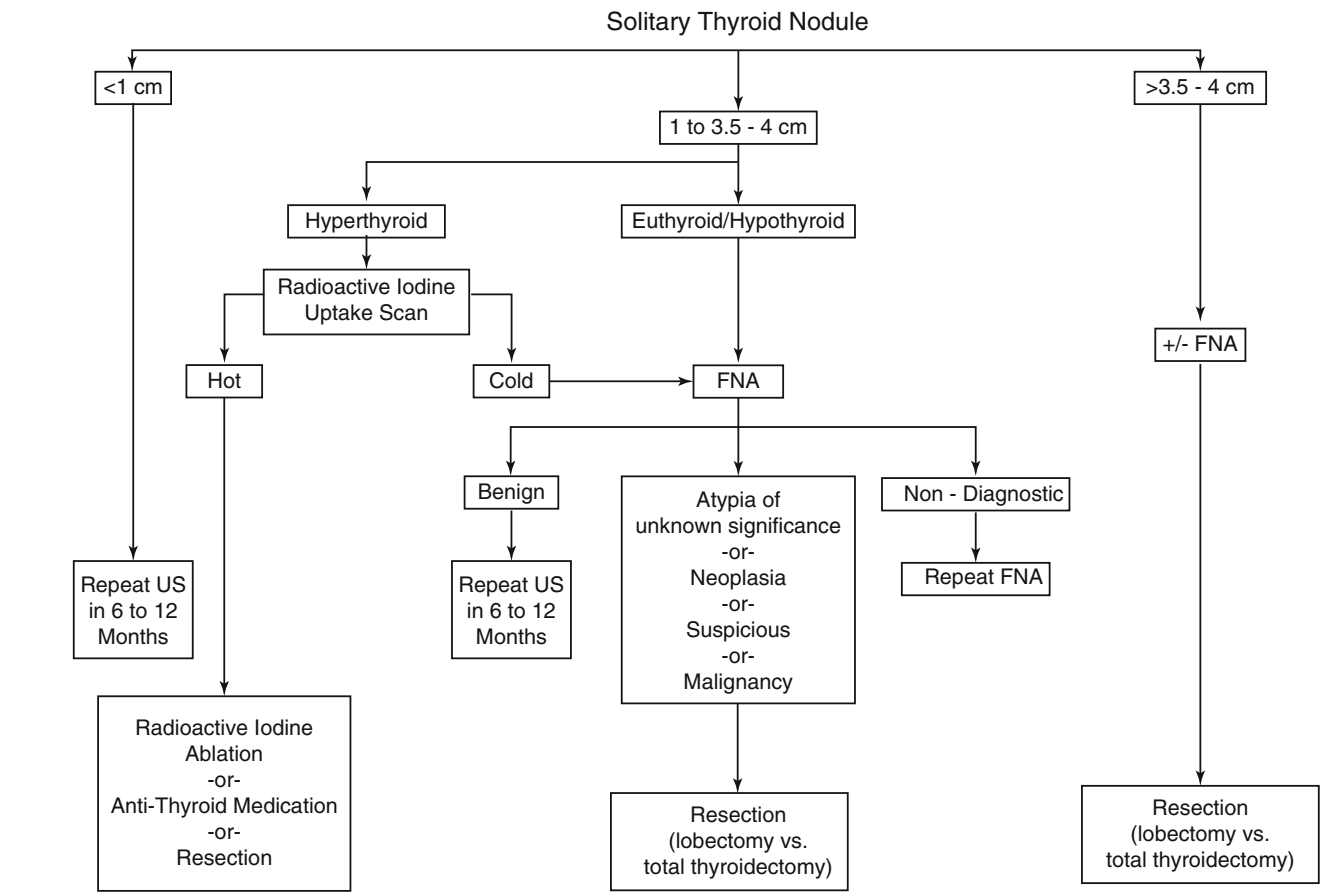

FIG. 22.1 Algorithm for evaluation and treatment of thyroid nodules. *FNA* fine-needle aspiration, *US* ultrasound

Hypothyroid or euthyroid patients require ultrasound-guided FNA of the nodule. Patients with a history of prior neck irradiation should be considered for an operation regardless of FNA result, as 20–50 % of patients in this scenario will be found to harbor thyroid cancer.

Fine-Needle Aspiration (FNA)

This diagnostic procedure requires an experienced endocrine surgeon, endocrinologist, or radiologist to perform the aspiration and an experienced cytopathologist to review the aspirate. It is a generally well-tolerated and facile procedure that can be performed in the office setting with ultrasound guidance and with a small-gauge needle. FNA can definitively diagnose papillary and medullary thyroid cancer; however, it cannot differentiate between benign and malignant follicular lesions. The cytopathology from FNAs has been traditionally classified as "inadequate," "benign," "indeterminate," or "malignant." However, the National Cancer Institute Thyroid Fine-Needle Aspiration Guidelines Committee recently proposed a modified classification. This new classification system (Table 22.1) is commonly referred to as the "Bethesda criteria."

TABLE 22.1. The National Cancer Institute's modified classification for FNA cytopathology.

Suggested category	Alternate category	Malignancy risk
Benign		<1 %
Atypia of undetermined significance	Indeterminate follicular lesions, R/O neoplasm, atypical follicular lesion, cellular follicular lesion	5–10 %
Neoplasm	Suspicious for neoplasm	20–30 %
Suspicious for malignancy		50–75 %
Malignant		100 %
Nondiagnostic	Unsatisfactory	

Treatment

A lesion that is "*benign*" can be observed with serial ultrasonography every 6 to 12 months. A change in size warrants repeat FNA or resection. A result of "*atypia of undetermined significance*" or "*neoplasia*" merits lobectomy or total thyroidectomy. Nodules that are "*suspicious for malignancy*" or "*malignant*" on FNA should be treated based on size. Patients with nodules less than 1 cm should undergo lobectomy or total thyroidectomy, whereas those more than 1 cm should

undergo total thyroidectomy with or without central lymph node dissection. A result of "*nondiagnostic*" necessitates a repeat FNA. The different types of thyroid malignancies and their features are discussed next.

Thyroid Malignancies

Thyroid malignancies include papillary (80 %), follicular (10 %), medullary (5–7 %), and anaplastic cancers (2–5 %). Papillary carcinoma is typically multifocal, is more commonly found in iodine-rich regions, spreads locoregionally and through lymphatics, and carries the best prognosis. Follicular cancers are often solitary, are more common in patients from iodine-poor regions, and spread hematogenously. Medullary thyroid carcinoma is derived from the parafollicular "C" cells that produce calcitonin. Seventy to eighty percent of cases are sporadic, with the remaining being inherited and associated with multiple endocrine neoplasia (MEN) II and familial medullary thyroid cancer syndromes. The patient and family should be screened for RET proto-oncogene due to an autosomal dominant pattern of inheritance. Anaplastic carcinoma typically displays rapidly progressing advancement and carries the poorest prognosis. When feasible, thyroidectomy is offered, with attempts at palliation (i.e., tracheostomy) and adjuvant therapies (i.e., external-beam radiation, chemotherapy) being employed for the remainder of cases.

Conclusion

As populations live longer and further advancements in imaging develop, clinicians will encounter an increasing number of patients with thyroid nodules. The possibility of malignancy underscores the importance of a thorough evaluation of these nodules. It is imperative to risk-stratify patients based on historical elements, to perform a complete head and neck exam, and to assess thyroid function. Imaging and FNA of nodules are then used to guide treatment decisions.

23
Cushing's Syndrome

Chung-Kay Koh and Yoojin Kim

Signs and Symptoms

Cushing's syndrome refers to a spectrum of diseases that result from prolonged exposure to excess glucocorticoids. Symptoms include fatigue, weight gain, insomnia, irritability, and menstrual irregularities. Signs include obesity, dorsocervical fat pad ("buffalo hump"), supraclavicular fullness, edema, thinning of skin, acne, and hirsutism. The most suggestive features of Cushing's include easy bruising, facial plethora, proximal muscle weakness, and reddish-purple striae. Associated diseases include diabetes, hypertension, and osteoporosis.

Cushing's syndrome can be classified by its cause: *adrenocorticotropic hormone (ACTH) dependent* or *ACTH independent* (Fig. 23.1).

ACTH-dependent causes are as follows:

1. Cushing's disease, from pituitary hypersecretion of ACTH (65–70 %)
2. Ectopic ACTH syndrome (nonpituitary hypersecretion of ACTH) (10–15 %)
3. Ectopic corticotrophin-releasing hormone (CRH) syndrome (nonhypothalamic tumor producing CRH causing pituitary hypersecretion of ACTH) (less than 1 %)
4. Iatrogenic administration of exogenous ACTH (less than 1 %)

ACTH-independent causes are as follows:

1. Iatrogenic administration of exogenous glucocorticoids (most common cause of Cushing's syndrome)
2. Adrenal adenomas and carcinomas (18–20 %)
3. Bilateral micronodular adrenal hyperplasia (less than 1 %)
4. Bilateral macronodular adrenal hyperplasia (less than 1 %)

Pseudo-Cushing's syndromes in which patients present with some features of Cushing's and mild biochemical hypercortisolism must be kept in mind; causes include alcohol use, depression, or obesity. Findings will resolve with correction of the underlying cause.

Diagnosis

The first step is to exclude, by taking a meticulous history, iatrogenic Cushing's syndrome due to chronic, exogenous glucocorticoid use or exposure (oral, inhaled, rectal, topical, and/or injected). If this has been ruled out, one of the following initial screening tests should be performed:

1. *24-h urine free cortisol*
A simultaneous 24-h urine creatinine should be measured to make sure the urine collection and volume is an adequate sample. A free urinary cortisol level greater than normal range for that laboratory should be repeated.

2. *Late night or midnight salivary cortisol*
The patient is given a salivette to chew on around 11 PM or midnight, when cortisol value nadirs. A salivary cortisol value greater than the upper limit of normal for that particular laboratory warrants repeat testing for confirmation.

3. *1 mg overnight or low-dose dexamethasone suppression test (LDDST)*
The patient is given 1 mg PO dexamethasone between 11 PM and 12 AM. An 8 AM serum cortisol is checked the following morning. A value less than 1.8 μg/dL has a high sensitivity for excluding Cushing's.

There are special caveats to keep in mind. For example, a 24-h urine cortisol is unreliable in a patient with renal failure. Various antiepileptic agents may increase the metabolism of dexamethasone making the dexamethasone

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_23, © Springer New York 2015

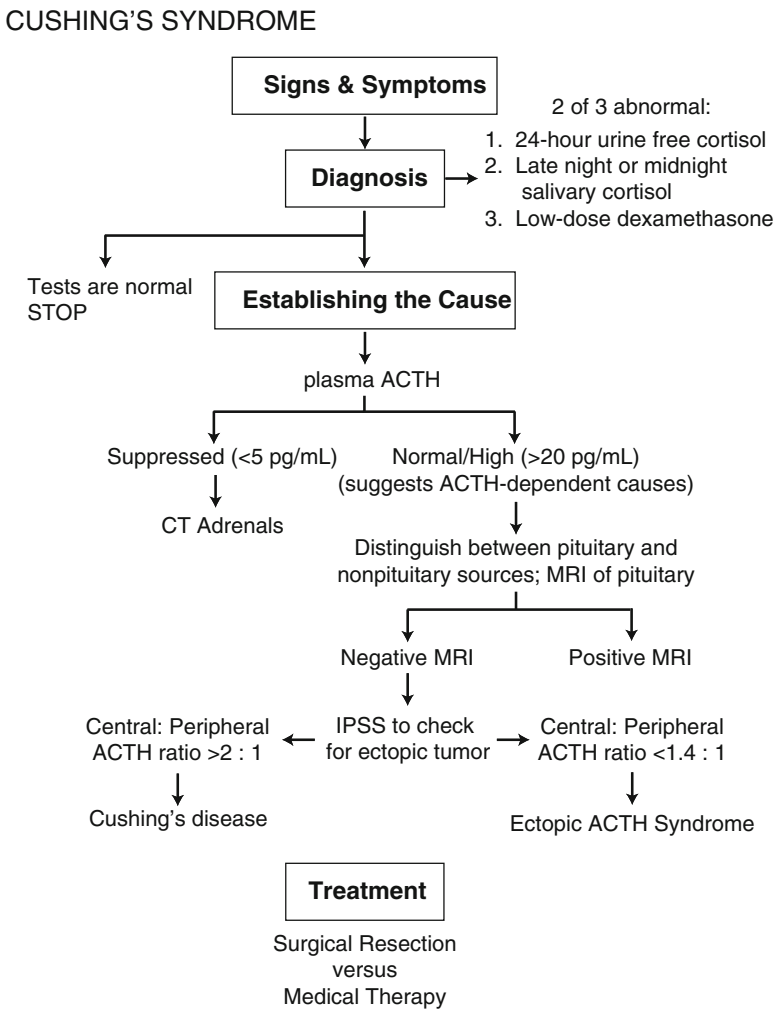

FIG. 23.1 Treatment algorithm for Cushing's syndrome

suppression test unreliable. Smoking increases the production of salivary cortisol.

Establishing the Cause

Once hypercortisolism has been confirmed biochemically by two of the three screening tests, the next step is to check a plasma ACTH level to distinguish between ACTH-dependent and ACTH-independent Cushing's syndrome.

A suppressed ACTH level (less than 5 pg/mL) points to the adrenals as the source. A computed tomography (CT) scan of the adrenals should be performed.

A normal to high ACTH level (usually greater than 20 pg/mL) suggests ACTH-dependent causes, which must now be distinguished between pituitary and nonpituitary sources. Since the most common of the ACTH-dependent cause is Cushing's disease or pituitary hypersecretion, a dynamic magnetic resonance imaging (MRI) of the pituitary is the next step. If the dynamic MRI is positive for a pituitary tumor, the source has been localized.

However, even if the MRI of the pituitary is negative for the source, there is still the possibility that a pituitary microadenoma versus an ectopic tumor is responsible. At this point in the diagnostic algorithm, various tests in the past have been used to attempt to localize the source: the high-dose dexamethasone suppression test, the metyrapone test, and CRH-releasing factor tests.

Currently, the most sensitive and specific test to distinguish Cushing's disease from an ectopic source is inferior petrosal sinus sampling (IPSS). IPSS uses selective catheterization and venous sampling of blood from the bilateral inferior petrosal sinuses, where blood from each half of the pituitary drains. ACTH levels are drawn simultaneously from the bilateral inferior petrosal sinuses and the periphery before and after corticotrophin-releasing hormone (CRH) administration. The central to peripheral ACTH ratios are compared. Under the premise that a pituitary adenoma still retains its responsiveness to CRH and an ectopic ACTH-producing tumor does not, *a central to peripheral ACTH ratio greater than 2:1 at baseline or greater than 3:1 after CRH administration has a 97 % sensitivity and 100 %*

specificity in diagnosing Cushing's disease. Also, a left to right central ACTH ratio greater than 2:1 can help to lateralize the location of the pituitary microadenoma in preparation for surgical resection. It has also been suggested that prolactin levels be drawn with each ACTH level to confirm that the sinuses were cannulated correctly.

If the central to peripheral ACTH ratio is less than 1.4:1, this suggests an ectopic source, although there are false-negative results. The etiologies of ectopic ACTH syndrome include: small cell lung cancer (50 %), lung carcinoids (10 %), pancreatic tumors (10 %), non-small cell lung cancer (5 %), thymic tumors (5 %), medullary carcinoma of thyroid (5 %), pheochromocytoma and related tumors (3 %), other carcinoids (2 %), and rare carcinomas of other organs (10 %). To detect these "occult" tumors, several different imaging modalities can be employed: CT or MRI of the chest, abdomen, and pelvis with 0.5 cm cross sections; 111Indium-labeled octreotide scan (as some neuroendocrine tumors express somatostatin receptors); and/or positron emission tomography (PET) scans.

Treatment

For the majority of cases, the treatment is surgical resection. This is true for adrenal adenomas, pituitary tumors leading to Cushing's disease, and ectopic ACTH syndrome with a localizing tumor. Even for adrenal carcinomas with metastatic disease, the primary tumor may be removed to improve the patient's response to postsurgical medical therapy. However, for cases in which the primary tumor cannot be found such as in ectopic ACTH syndrome, bilateral adrenalectomies are employed to improve hypercortisolism. Medical therapies include: adrenolytics (ketoconazole, mitotane, metyrapone), ACTH secretion inhibitors (cabergoline, octreotide), and glucocorticoid receptor antagonists (mifepristone). Medical therapies may be used to optimize patients in preparation for surgery or postsurgically due to persistence or recurrence of the Cushing's syndrome.

24
Hyperthyroidism

Tricia A. Moo-Young

History

Hyperthyroidism or thyrotoxicosis is the clinical state of excessive serum concentrations of thyroxine (T_4), triiodothyronine (T_3), or both coupled with suppressed levels of thyroid-stimulating hormone (TSH). In the United States, hyperthyroidism has an estimated prevalence of 1.2 %. Based purely on the biochemical profile, hyperthyroidism is identified as either "overt" or "subclinical." *Subclinical* hyperthyroid patients have a suppressed TSH with normal levels of T_4 or T_3. Typically patients have no symptoms of hyperthyroidism at clinical presentation. *Overt* hyperthyroidism is defined by a suppressed TSH with elevated levels of T_4 or T_3 . These patients typically exhibit signs and symptoms of their condition but can be entirely asymptomatic. Clinical signs and symptoms include nervousness, palpitations, flushing, weight loss, increased sweating, difficulty concentrating, and emotional liability. In women the menstrual cycle is often irregular or absent. Patients with an exaggerated state of hyperthyroidism can present with the life-threatening condition known as "*thyroid storm*." Signs and symptoms include fever, cardiac arrhythmias, circulatory shock, multiorgan failure, and are invariably fatal if left untreated (Fig. 24.1).

Physical Examination

Physical signs of hyperthyroidism include tachycardia, elevated blood pressure, possible thyroid enlargement and or bruit, skin changes, and tremor. Additional physical features can be associated with specific causes of hyperthyroidism.

Diffuse Enlargement

Graves' disease is the most common cause of hyperthyroidism in all age groups within the United States. It is an autoimmune disorder caused by a stimulating antibody toward the thyrotropin (TSH) receptor. The result is unregulated induction of TSH production and downstream production of T_3 and T_4. Patients may have a diffusely enlarged thyroid with a prominent bruit upon auscultation. The gland can be tender to touch, but a painful goiter should prompt consideration of subacute thyroiditis. Proptosis, lid lag, and periorbital edema are the characteristic physical findings of Graves' orbitopathy (GO); these findings are found in one-third of all Graves' patients. Additional physical findings include soft tissue enlargement of the fingers with clubbing known as *thyroid acropachy* and *thyroid pretibial myxedema*.

Nodular Enlargement

Multinodular toxic goiters (MNGs) are associated with unregulated autonomous synthesis of thyroid hormones T3 and T4 from nodules within the thyroid gland. It is the second most common cause of hyperthyroidism in the United States behind Graves' disease. Among older patients, toxic MNG is the most common cause of hyperthyroidism. The majority of thyrotoxic nodules are caused by somatic mutations of the TSH receptor. The natural history of a multinodular goiter involves variable growth of the individual nodules; enlargement can cause aerodigestive compression or may be undetectable on physical exam if there is large substernal component. When enlargement is significant, simply elevating a patient's arms above the head can precipitate a classic "*Pemberton sign*" evidenced by marked facial plethora. This is caused by mechanical compression of the neck veins by narrowing of the thoracic inlet with arm elevation.

Toxic Adenomas (Plummer's Disease)

Toxic adenomas (Plummer's disease) represent a single hyperfunctioning nodule within the background of a suppressed multinodular goiter or normal thyroid gland. These

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_24, © Springer New York 2015

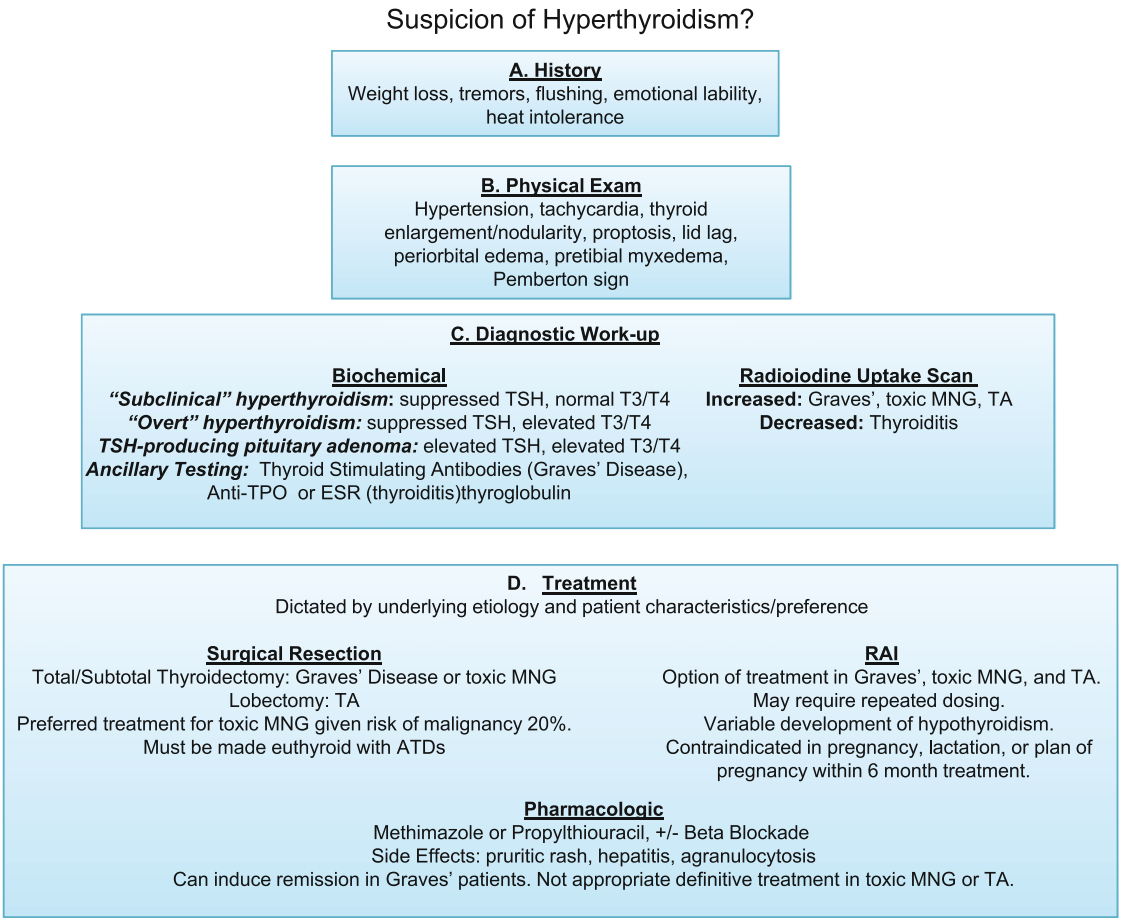

FIG. 24.1 Treatment algorithm for hyperthyroidism. *TPO* thyroid peroxidase, *MNG* multinodular goiter, *TA* toxic adenoma, *ESR* erythrocyte sedimentation rate, *ATD* antithyroid drugs

nodules are almost always benign. A somatic mutation similar to that found in MNG is accountable for the hyperthyroid state in 20–80 % of toxic nodules studied.

Other Causes of Hyperthyroidism

Other rare causes of hyperthyroidism include thyrotoxic phase of subacute thyroiditis, pituitary resistance to thyroid hormone, metastatic thyroid cancer, and struma ovarii. Iatrogenic causes include factitious hyperthyroidism and pharmacologic agents such as amiodarone that inhibit T4 to T4 conversion.

Diagnosis

Serum TSH measurement is the best initial step in the evaluation of patients with suspected hyperthyroidism. Levels should be low regardless of whether the patient has overt or subclinical hyperthyroidism. It should, however, be kept in mind that older patients will usually have a higher normal TSH level compared with their younger counterparts. If the TSH level is suppressed, the next biochemical testing should include free T_4 and total or free T_3 levels. Graves' disease patients typically have T_3 thyrotoxicosis rather than pure T_4. Once the diagnosis of hyperthyroidism is confirmed, further testing to determine the cause can be performed. Graves'

disease measurement of thyroid-stimulating immunoglobulin (TSI) can be done to help confirm the diagnosis. This is especially useful during the last few weeks of pregnancy where high levels of TSI in maternal blood are associated with increased incidence of transient hyperthyroidism in newborns. Antithyroid peroxidase (TPO) antibodies are measured to confirm a suspected diagnosis of autoimmune thyroid disease and to assess the risk for drug-induced thyroid dysfunction.

Radioiodine uptake scan (RAIU) is indicated when the underlying cause of hyperthyroidism is not yet established or when nodularity is present. In Graves' disease and toxic MNG, the distribution of RAIU is diffuse and nodular, respectively. Alternatively a toxic adenoma will show focal uptake known as "hot nodule" with a suppressed background of normal thyroid parenchyma. Decreased uptake in a nodular distribution can represent a "cold nodule," which has an increased malignant potential. Such findings should be investigated further with appropriate imaging and biopsy where indicated. The RAIU will be suppressed in the presence of subacute or postpartum thyroiditis. *RAIU scans are contraindicated during pregnancy* and thus the diagnosis is made clinically and with laboratory testing. In patients with suspected iatrogenic or factitious hyperthyroidism, a low serum thyroglobulin level confirms the diagnosis. A patient in whom the TSH, T_3, and T_4 levels are all elevated should have imaging of the pituitary gland to determine the presence of TSH-producing pituitary adenoma.

Treatment

Treatment is based on the underlying clinical cause. The three major treatment options include antithyroid drugs (ATDs), radioiodine ablation (RAI), and surgical resection. Choice of definitive treatment is typically made during a discussion between the physician and patient regarding such matters as the logistics, benefits, costs, and potential side effects of each treatment option. *Antithyroid drugs* include methimazole or propylthiouracil (PTU); side effects include pruritic rash, hepatic dysfunction, and agranulocytosis. *Methimazole* is given at an initial dose of 20–40 mg daily and PTU is 100 mg every 8 h. Symptomatic management of palpitations, tachycardia, and emotional lability is accomplished through the use of beta-adrenergic blockade. ATDs are contraindicated in hyperthyroid patients exhibiting major adverse reactions to the medications. *RAI* is typically given either as a fixed dose or a calculated dose based on the activity and size of the gland. RAI is contraindicated in women planning pregnancy within 4–6 months of treatment, active pregnancy, lactating women, and a patient with suspicion of thyroid cancer. *Surgical options* include lobectomy in nodular disease and subtotal or total thyroidectomy in MNG or Graves' disease. Complications in either procedure include transient or permanent hypoparathyroidism and/or recurrent laryngeal nerve injury. *Prior to surgery all hyperthyroid patients should be rendered euthyroid with ATDs* to avoid precipitating a thyroid crisis during or after the procedure. Additionally to help reduce blood loss related to the hypervascularity of the gland prior to surgery, *patients are given potassium iodide* (five drops given three times a day for 5 days prior to surgery) to induce vasoconstriction of the gland. Surgery is not an option for any patient considered to be a poor surgical candidate (i.e., comorbid conditions, elderly, prior neck radiation/surgery).

Graves Disease

Management of patients with Graves' disease can be accomplished by any of the three available modalities: RAI, ATDs, or thyroidectomy. Methimazole is typically the first drug of choice in patients with Graves' disease, *except in pregnancy and thyroid storm where PTU is preferred*. In the United States, the rate of remission in Graves' disease patients is approximately 20 % after 12–18 months of treatment with antithyroid drugs. Favorable predictors of remission include mild disease, small goiters, and negative antibody levels. The optimal therapeutic strategy for Graves' disease patient with eye symptoms remains controversial. Overall the use of ATDs or thyroidectomy does not alter the clinical course of GO. RAI, however, can cause de novo development or progression of GO in Graves' patients. This can typically be prevented with corticosteroid pretreatment. Surgical resection for Graves' disease patients includes total or subtotal thyroidectomy. Total thyroidectomy versus subtotal has a 0 and 8 % risk of recurrent hyperthyroidism, respectively. Finally, Graves' patients with thyroid nodules should have an appropriate malignancy workup prior to initiating any definitive therapy.

Toxic Multinodular Goiter

According to the 2011 American Thyroid Association Guidelines, patients with toxic MNG or TA should be treated with either RAI or thyroidectomy. In toxic MNG patients surgical resection has a <1 % risk of treatment failure, whereas RAI has a 20 % risk of need for repeat treatment. RAI can reduce goiter volume by as much as 40 % within the first two years following treatment. The prevalence of hypothyroidism posttreatment is 100 % in surgical resection versus 3 % at 1 year and 64 % at 24 years in RAI patients. With RAI the risk of posttreatment hypothyroidism is higher for patients requiring repeat treatments and patients less than 50 years of age. Increasing evidence now suggests a nearly 20 % risk of concomitant thyroid malignancy in patients with toxic MNG. For this reason, many clinicians favor surgical resection over RAI.

Toxic Adenoma

Toxic adenoma patients have 99 % treatment success following surgical resection compared to 82–94 % in RAI-treated patients. RAI is given at either a fixed total dose or an activity level based on nodule size. Establishment of a euthyroid state is typically immediate following surgical resection for TA and in RAI within three months for 75 % of patients. Reduction in the nodule volume averages around 35 % at 3 months following treatment. The development of postsurgical hypothyroidism is only 2.3 % with surgery. Among patients treated with RAI, the prevalence is variable depending on adequacy of TSH suppression and presence of antithyroid antibodies posttreatment.

25
Hypercalcemia

Malini D. Sur and Kaare J. Weber

Introduction

Hypercalcemia is a relatively common disorder with significant clinical impact. Mild chronic hypercalcemia can produce low-grade but long-lasting symptoms that surreptitiously interfere with routine daily activities. Severe acute hypercalcemia is fatal if not recognized and treated immediately.

Biology

Serum calcium (Ca) levels are primarily regulated by parathyroid hormone (PTH) and calcitriol. A slight decrease in the plasma concentration of ionized Ca stimulates chief cells of the parathyroid glands to secrete PTH, a peptide with a half-life of 2–4 min. In the bone, PTH activates osteoclasts to dissolve bone matrix and liberate Ca. In the distal nephron it increases Ca ion reabsorption from the urine, and in the proximal nephron it enables production of calcitriol—the active form of vitamin D. Calcitriol promotes intestinal Ca absorption, renal tubular Ca reabsorption, and activation of osteoclasts by osteoblasts. These pathways act in concert to increase serum Ca and decrease urinary Ca. PTH is counteracted by calcitonin, a hormone derived from parafollicular cells of the thyroid. Calcitonin inhibits intestinal Ca absorption, renal tubular Ca reabsorption, and osteoclast activation, but its effects are minor compared to PTH.

Clinical Presentation

Hypercalcemia is typically discovered incidentally on routine blood work in seemingly asymptomatic patients. In these cases, particularly when serum Ca levels are above 11 mg/dL, a detailed history may elucidate vague chronic symptoms often involving multiple systems (Fig. 25.1). These include depression, fatigue, polyuria, renal stones, constipation, abdominal pain, and muscle aches. Hypercalcemic crisis occurs in patients with serum Ca typically above 14 mg/dL and presents with constitutional, gastrointestinal, and cardiac symptoms. All patients with hypercalcemia should be asked about underlying renal disease and cancer. A medication history may reveal use of thiazides or lithium. Overconsumption of Ca, vitamin D, or vitamin A should be excluded. Family history should look for multiple endocrine neoplasia syndromes or early onset hypercalcemia. Though enlarged parathyroids are rarely palpable, a thorough head and neck exam will help exclude any concomitant conditions, such as thyroid nodules, that should be addressed preoperatively. The remainder of the physical exam should focus on signs of occult cancer.

Laboratory Investigations

Because nearly half of serum Ca is bound to albumin, serum Ca levels must be corrected for the amount of albumin present. Pseudohypercalcemia can occur in dehydrated patients or those with multiple myeloma who have a circulating paraprotein that also binds Ca. Patients that have true hypercalcemia with serum Ca below 14 mg/dL and mild to no symptoms can undergo an outpatient workup, which begins with measurements of intact PTH and phosphate levels. Those with acute, severe symptoms or serum Ca above 14 mg/dL should be referred to the emergency department and stabilized according to the guidelines outlined below.

Hypercalcemic Crisis

Hypercalcemic crisis is a life-threatening condition requiring immediate diagnosis and treatment. The most common etiology is malignancy-associated hypercalcemia (MAH), although medications, granulomatous diseases such as sarcoidosis and tuberculosis, and undiagnosed primary hyperparathyroidism (HPT), particularly with underlying

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_25, © Springer New York 2015

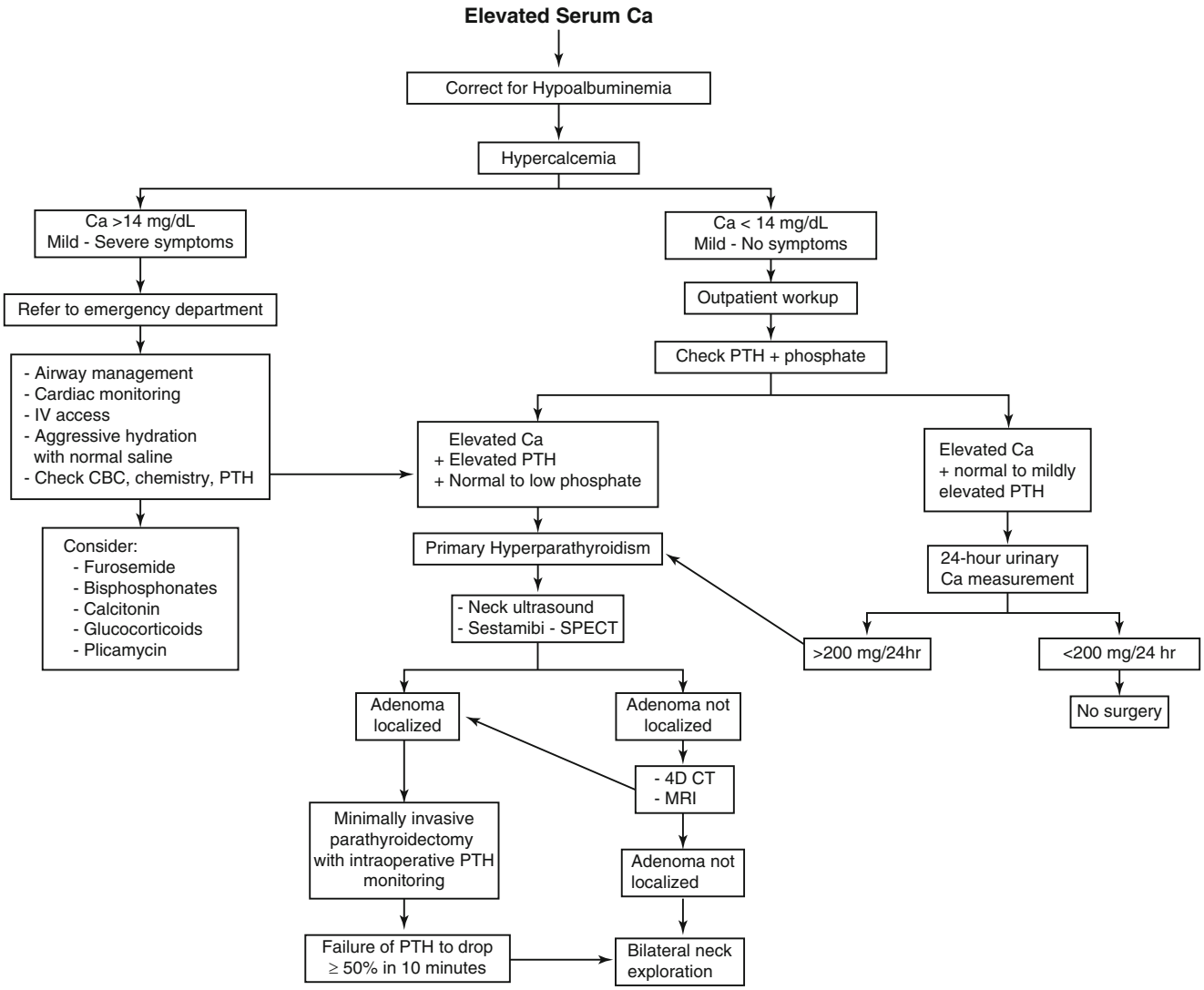

FIG. 25.1 Treatment algorithm for hypercalcemia. *Ca* calcium, *PTH* parathyroid hormone, *IV* intravenous, *CBC* complete blood count, *SPECT* single photon emission computed tomography, *CT* computed tomography, *MRI* magnetic resonance imaging

parathyroid carcinoma, must also be considered. Symptoms include nausea, vomiting, fatigue, weakness, lethargy, and arrhythmias. Even with mild symptoms, a serum Ca level above 14 mg/dL should be treated expeditiously. Therapy begins with airway assessment, cardiac monitoring, establishment of intravenous access, and aggressive hydration with normal saline to maintain a urine output above 100 cc/h. This treats dehydration, which can both be the cause and result of hypercalcemia. A complete blood count, full chemistry panel, and PTH level are sent at this time. Furosemide can be used to induce further diuresis and Ca excretion. Caution must be used to avoid overdiuresis. Adjunct agents include bisphosphonates, calcitonin, glucocorticoids, and plicamycin. Bisphosphonates inhibit osteoclasts and bone resorption and can potentially normalize serum Ca levels within 2–3 days. Subcutaneous calcitonin immediately inhibits osteoclast activity, but tachyphylaxis prevents sustained results. Glucocorticoids block cytokines affecting bone resorption, increase Ca excretion, and decrease intestinal Ca absorption. Plicamycin is an inhibitor of osteoclast RNA synthesis. In cases of severe, refractory hypercalcemic crisis, hemodialysis may be necessary. If PTH levels are elevated, primary HPT can be treated with an urgent parathyroidectomy once the patient is adequately resuscitated, electrolytes are normalized, and localization studies are completed.

Hypercalcemia with Elevated PTH

Primary Hyperparathyroidism

Elevated serum Ca, elevated PTH, and normal to decreased phosphate levels are consistent with primary HPT. A solitary parathyroid adenoma is responsible for nearly 85 % of cases. Parathyroid hyperplasia accounts for 10–15 % of cases while multiple adenomas are found in less than 5 %. Less than 1 % of primary HPT is due to an underlying parathyroid carcinoma. Once primary HPT is diagnosed, accurate preoperative localization of an occult adenoma allows a focused, or minimally invasive, parathyroidectomy (MIP) via a small cervical incision with dissection limited to the suspected abnormal side. Given that no one imaging modality is reliably sensitive for the identification of an adenoma, most endocrine surgeons use at least two scans to improve the odds of successful localization. All patients with primary HPT should first undergo *neck ultrasonography* (US) to look for an oval, hypoechoic or anechoic mass posterior to the thyroid gland, which would suggest a likely parathyroid adenoma. Concomitant thyroid disease can also be excluded with this study, and intrathyroidal parathyroid glands can be identified. Scintigraphy is typically obtained next using *technetium-99 sestamibi* with single photon emission computed tomography (sestamibi-SPECT), which identifies most hyperfunctioning parathyroid tissue but is less sensitive for multiglandular disease. When both studies have a positive, concordant result, the positive predictive value approaches 100 %. More recently, 4D computed tomography has been used as an alternative to sestamibi-SPECT or as an adjunct when other studies are equivocal. Magnetic resonance imaging can also be used when sonography and scintigraphy are unrevealing.

Once an adenoma is definitively visualized on imaging, the surgeon can proceed with MIP with *intraoperative PTH monitoring*. The parathyroid glands are typically positioned symmetrically, with the low glands lying anterior to the recurrent laryngeal nerve (RLN) and the upper glands lying posterior to the RLN as it enters the cricothyroid muscle. Aberrant glands may also be found cephalad to the superior pole of the thyroid, along the great vessels of the neck in the tracheoesophageal groove, within the thymic tissue, within the thyroid parenchyma itself, or in the mediastinum. It is important to avoid excess trauma to prevent devascularization and rupture. The latter may lead to parathyromatosis, or seeding of the parathyroid tissue, which raises the risk of recurrent hyperparathyroidism. If an aberrant-appearing gland is confirmed in the predicted location, it should be resected. *Failure of the PTH level to fall by at least 50 % within 10 min after excision should prompt a bilateral exploration. In addition, if the gland found in the predicted location appears completely normal, the remaining glands should be inspected*. If the PTH falls appropriately with removal of the adenoma, the incision is closed after hemostasis is achieved.

If preoperative imaging fails to identify an adenoma, the surgeon should perform bilateral neck exploration with PTH monitoring. If all four parathyroid glands are identified and hyperplasia is confirmed by frozen section biopsy, 3½ glands are excised. If only three glands can be identified, hemithyroidectomy can be considered on the side of the missing gland, though ideally intrathyroidal parathyroids are identified preoperatively using ultrasound or fine needle aspiration (FNA) with measurement of aspirate PTH levels.

Familial Hypocalciuric Hypercalcemia (FHH)

In patients with only a mildly elevated serum Ca and a normal to mildly elevated PTH, FHH should be considered. FHH is the result of a silencing mutation in the Ca-sensing receptor of the parathyroid glands and kidneys. Measurement of 24-h urine Ca will distinguish FHH from primary HPT. In FHH, daily Ca excretion will generally be less than 200 mg. FHH typically has a benign natural history and cannot be cured with surgery. *Parathyroidectomy should thus be avoided in patients with FHH*.

Hypercalcemia with Suppressed PTH

Malignancy-Associated Hypercalcemia

Malignancy-associated hypercalcemia (MAH) should be considered in any patient with new symptoms of hypercalcemia with serum Ca above 14 mg/dL, low phosphate levels, and suppressed PTH. Blood work may also show anemia and increased alkaline phosphatase levels and erythrocyte sedimentation rate. MAH affects up to 25 % of cancer patients and is a poor prognostic indicator. In most cases, PTH-related protein (PTHrP) secreted by tumor cells within bony metastases and extraskeletal sites stimulates osteoblasts to express receptor activator of nuclear factor-KB ligand (RANKL), which in turn activates osteoclasts. The most common cancers associated with PTHrP-mediated MAH are squamous cell lung carcinoma, breast carcinoma, and renal cell carcinoma. MAH is also seen in multiple myeloma and some cases of lymphoma, where different cytokines are involved in stimulating osteoclasts to resorb bone. Conventional treatment of MAH consists of bisphosphonates and glucocorticoids. Denosumab, a monoclonal antibody against RANKL, has emerged as a new tool to help prevent skeletal-related events in patients with solid tumors and bony metastases.

26
Insulinoma

Steven A. Jong

Introduction

Insulinomas are the most common functional neuroendocrine tumor of the pancreas and can occur sporadically (90 %) or as part of the multiple endocrine neoplasia type I (MEN-I) syndrome (10 %). Accounting for 70–80 % of all pancreatic neuroendocrine neoplasms, the estimated incidence has increased to four occurrences per one million people annually. Due to the rarity of these tumors, however, many patients are initially misdiagnosed in clinical practice, and the median duration of symptoms prior to diagnosis is 3–5 years. The great majority of insulinomas are solitary, benign, and distributed equally throughout the head, body, and tail portions of the pancreas. Typically they are small tumors with greater than 80 % being less than 2 cm in diameter. Over 90 % of insulinomas are non-metastatic at the time of presentation and can be surgically cured. Surgical excision through either tumor enucleation or anatomic pancreatic resection is the treatment of choice and offers the only opportunity for cure. The type of resection depends on tumor size, location, and several pathologic characteristics of the tumor. Laparoscopic management of insulinoma is feasible and safe for tumors located in the body or tail of the pancreas. Open surgery combined with intraoperative ultrasonography is recommended for proximal pancreatic tumors or for patients with multiple insulinomas (10 %) commonly seen in association with inherited endocrine syndromes. Insulinoma is the second most frequent functioning pancreatic endocrine tumor in MEN-I patients with a prevalence of 10–20 %. Aggressive behavior and malignancy is rare and seen in only 5–10 % of patients who demonstrate distant metastasis to regional lymph nodes, liver, or bone. This clinical presentation is associated with a median survival of less than 2 years.

Diagnosis

The diagnosis of insulinoma begins with a clinical suspicion arising from *symptomatic hypoglycemia* (Fig. 26.1). These symptoms can take the form of *neurologic changes*, which can include altered mental state, confusion, visual disturbances, generalized weakness, and abnormal behavior. Hypoglycemia also results in catecholamine release precipitating adrenergic *sympathetic nervous system* activation and associated sweating, anxiety, and palpitations. Patients also commonly gain weight as they increase their carbohydrate intake to decrease the frequency and severity of their symptoms. These hypoglycemic symptoms, often precipitated by fasting and exercise, prompt the immediate measurement of plasma glucose levels, which are often less than 45–50 mg/dL. Symptomatic hypoglycemia is confirmed and these *symptoms disappear with the administration of carbohydrate*. These three criteria, known as *Whipple's triad*, are met and plasma insulin levels are found to be inappropriate or elevated (≥3–10 μ (mu) U/mL) and the insulin-to-glucose ratio is greater than 0.3 at the time of hypoglycemia. Elevated C-peptide (≥200 pmol/L) and proinsulin levels (≥5 pmol/L) confirm endogenous hyperinsulinism and, combined with the absence of plasma sulfonylurea, exclude the surreptitious use of insulin or oral hypoglycemic agents. The traditional 72-h fast is rarely needed or tolerated by these patients as 97 % will exhibit symptomatic hypoglycemia within 48 h of a supervised fast as plasma levels of glucose, insulin, and proinsulin, drawn every 6 h, are diagnostic of insulinoma. Once the patient develops neuroglycopenic symptoms, serum glucose, insulin, and C-peptide levels are obtained and the fast is terminated. Intravenous glucose is administered and the symptoms improve and disappear.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_26, © Springer New York 2015

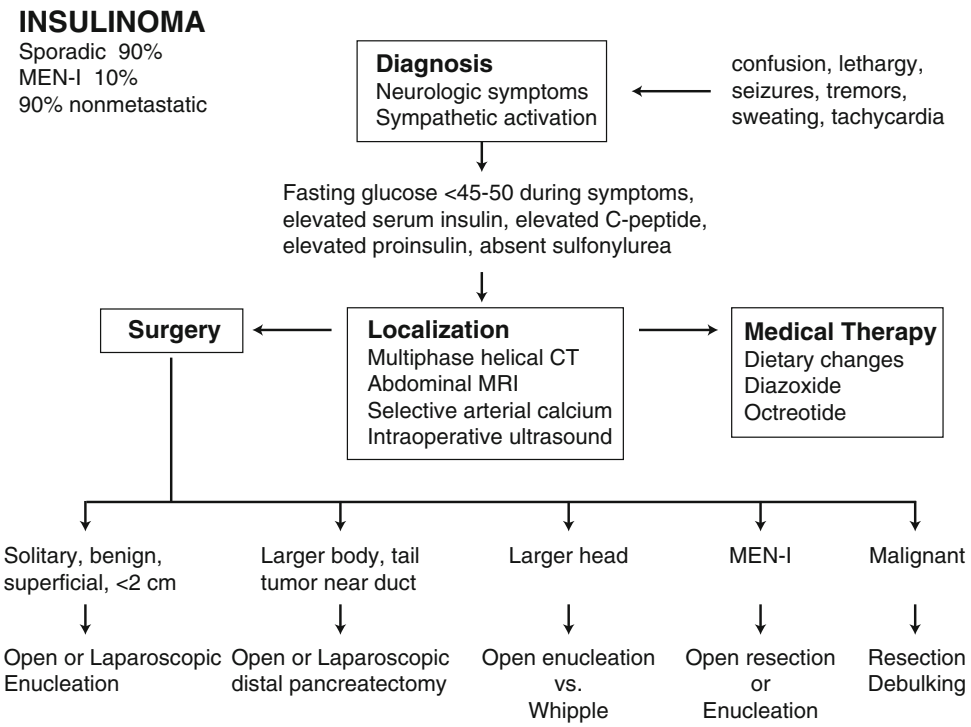

FIG. 26.1 Treatment algorithm for insulinoma. *MEN-I* multiple endocrine neoplasia type I, *CT* computed tomography, *MRI* magnetic resonance imaging

Localization

Once the diagnosis of insulinoma is confirmed, preoperative imaging studies are advised to exclude metastatic disease and to accurately determine the appropriate and optimal type of surgical resection. Despite many options for imaging these tumors, single or combination studies fail to identify the tumor in 10–30 % of patients. Preoperative transabdominal ultrasound (US), if performed by an experienced ultrasonographer, can be useful, and endoscopic US is an effective tool to localize tumors located in the pancreatic head and uncinate process. Multiphase helical computed tomography (CT) with contrast for pancreatic perfusion using thin, 2 mm slices or abdominal magnetic resonance imaging (MRI) appear to be 70–90 % successful in localizing the tumor and may also demonstrate metastases if present. Intra-arterial calcium stimulation with hepatic vein sampling can pinpoint the area of the pancreas containing the insulinoma in patients with negative three-dimensional imaging or when initial abdominal and pancreatic exploration fails to cure the patient. Calcium, a known secretagogue for insulin secretion from the insulinoma, is injected sequentially into the catheterized gastroduodenal, superior mesenteric, and proximal and distal splenic arteries combined with timed measurement of insulin from catheterized right and left hepatic veins. Elevated hepatic vein insulin levels after injection of the gastroduodenal, superior mesenteric, or splenic artery would localize the insulinoma to the head, neck, or tail of the pancreas, respectively. A tumor blush, if seen during these arterial injections, can also assist in the localization process. Insulinomas are infrequently detected on preoperative octreotide scanning as they often lack somatostatin receptors, and invasive studies like pancreatic angiography and transhepatic portal venous sampling have been largely abandoned. Intraoperative US (IOUS) of the pancreas, introduced in 1981, identifies non-palpable insulinomas in 85–95 % of patients and is still considered by some to be the only localizing study necessary for these patients. IOUS can be used during both open and laparoscopic explorations to identify both solitary and multiple tumors and is a valuable operative tool to determine the proximity of the tumor to the pancreatic or biliary ductal system.

Medical Treatment

Dietary modifications and pharmacologic agents to control the symptoms of hypoglycemia and hyperinsulinemia are initiated after a failed abdominal exploration, for patients who are poor surgical candidates, those awaiting surgery, or as palliative treatment for metastatic disease. Patients are encouraged to eat small frequent meals throughout the day and at night to prevent symptoms. The initial drug of choice is diazoxide, which enhances glycogenolysis and stimulates

α(alpha)-adrenergic receptors to directly inhibit insulin release from the pancreatic β(beta)-cells. A dose range of 150–400 mg/day in divided doses titrated to control symptoms is effective in 50–60 % of patients, but side effects such as sodium retention, peripheral edema, gastrointestinal (GI) symptoms, and hirsutism can limit its usefulness. Somatostatin analogs octreotide and lantreotide, Dilantin, verapamil, glucocorticoids, and glucagon are other medical options for these patients.

Surgical Treatment

Most insulinomas are benign and curable by surgical excision in 90–95 % of patients with an operative mortality of 1–2 %. The entire abdomen and pancreas is thoroughly explored to identify the tumor and evaluate the presence or absence of malignant and/or metastatic disease. The insulinoma is commonly located intraoperatively with bidigital palpation and IOUS in 85–95 % of patients. Enucleation of the tumor and capsule is the procedure of choice if the tumor is on the surface of the pancreas and 2–3 mm away from the main pancreatic duct. Insulinomas without a clear capsule or those deeply embedded in the pancreas and adjacent to the pancreatic or biliary duct require formal pancreatic resection. Fixed, hard, lesions infiltrating into other surrounding structures and causing ductal dilatation suggest malignancy and mandate segmental pancreatectomy or, rarely, pancreaticoduodenectomy using standard oncologic principles to achieve negative margins and remove as much metastatic disease as possible. *Blind distal pancreatectomy for tumors not identified during exploration is not advisable given the equal distribution of these tumors throughout the pancreas*. Current localization options and advancements have made this procedure obsolete and unnecessary. Multiple insulinomas, commonly seen in patients with the MEN-I syndrome, are optimally treated with distal pancreatectomy to the level of the portal vein and synchronous enucleation of individual tumors in the pancreatic head. Abdominal exploration and tumor enucleation or segmental pancreatectomy for solitary insulin-secreting tumors can also be successfully performed laparoscopically, using laparoscopic ultrasonography to localize the tumor and utilizing HandPort placement, if needed, to facilitate palpation and resection of the pancreas. Severe adhesions, technical difficulty, extensive intra-abdominal fat, or splenic vein/capsule bleeding can all necessitate conversion from laparoscopic to open resection. Insulinomas that cannot be localized laparoscopically, are deeply immersed in the pancreatic parenchyma, or have a close relationship to the main pancreatic duct may also require open conversion. Intraoperative insulin measurement has been reported but not widely used.

Complication rates for both enucleation and formal pancreatic resection, with either an open or laparoscopic procedure, are similar at 15–45 % and often originate from a pancreatic duct disruption. This leakage results in pseudocyst formation, abdominal infection, abscess, fistula, and temporary delayed gastric emptying. These patients are often successfully managed with conservative measures including total parenteral nutrition, octreotide, CT-guided percutaneous drainage, antibiotics, and intestinal motility stimulants. Endoscopic pancreatic duct stenting and abdominal reexploration are rarely necessary, and intra-abdominal bleeding, atelectasis, pulmonary infection, and pulmonary embolism are other commonly reported complications.

Malignancy and Metastatic Disease

A malignant insulinoma is often identified by the discovery of metastatic disease during preoperative localization or during postoperative imaging. Operative findings of metastasis or local tumor invasion seen during surgical exploration and resection will also confirm malignancy. Common sites of metastases include the liver, regional lymph nodes, bone, and peritoneal surfaces. The clinical course of metastatic disease is variable as some patients may have few if any symptoms for years after diagnosis. Patients with severe symptoms associated with refractory hyperinsulinemia from recurrent or metastatic disease require pharmacologic control of insulin secretion to treat the debilitating and life-threatening symptoms of hypoglycemia. Most of these primary tumors are large with extensive metastases. Useful agents to control both preoperative and postoperative symptoms include diazoxide, glucagon, octreotide, and corticosteroids. Aggressive surgical debulking when possible, radiofrequency ablation, hepatic perfusion/embolization, peptide-receptor radionuclide therapy, biotherapy with interferon or long-acting octreotide, and chemotherapy with streptozotocin, doxorubicin, or 5-fluorouracil have all been utilized with varied success, but the prognosis remains poor with a median survival of 2 years. Everolimus and sunitinib have been recently introduced as targeted therapies for patients with metastatic pancreatic neuroendocrine tumors. Hepatic transplantation for unresectable liver metastasis with no extrahepatic involvement is rarely if ever performed.

27
Gastrinoma

Elizabeth C. Gwinn

Background

Gastrinoma is the second most common islet cell tumor of the pancreas and the most common symptomatic malignant endocrine tumor of the pancreas. About half of gastrinomas occur in the duodenum. Gastrinomas are associated with a syndrome, first described in 1955 by Drs. Zollinger and Ellison, characterized by severe peptic ulcer disease, hypersecretion of gastric acid, and a non-beta islet cell tumor of the pancreas (Fig. 27.1). This tumor is now understood to be a gastrin-secreting duodenal or pancreatic neuroendocrine tumor—or gastrinoma—and it is the cause of peptic ulcer disease in 0.1–1 % of patients. Zollinger-Ellison syndrome (ZES) occurs sporadically in 75–80 % of cases and as part of multiple endocrine neoplasia I in the rest. There are other causes of hypergastrinemia associated with acid hypersecretion, such as retained gastric antrum syndrome, chronic gastric outlet obstruction, chronic renal failure, significant small bowel resection, and gastric G-cell hyperplasia; however, these are rare. The major morbidity and mortality of gastrinomas was formerly related to complications of severe peptic ulcer disease. The development of H2 antagonists and proton pump inhibitors has produced a significant decrease in morbidity and mortality.

Presentation

Gastrinomas generally present with clinical symptoms due to hypersecretion of gastric acid causing refractory peptic ulcer disease. Gastrin has a trophic action on parietal cells and gastrin stimulates parietal cells via the release of histamine. Patients present with epigastric pain and heartburn; 90 % have peptic ulcers. Ulcers can be located in the duodenum, jejunum, or ileum and can cause symptoms such as nausea and vomiting, bleeding, or even perforation. A secretory diarrhea that is stopped by nasogastric aspiration of gastric secretions can also occur. Gastrinoma and ZES should be excluded in patients with intractable peptic ulcers, severe esophagitis, and persistent secretory diarrhea. In patients with multiple endocrine neoplasia type 1 (MEN-1) syndrome, elevated calcium, elevated parathyroid hormone (PTH) levels, or a pituitary tumor may also be present. Hyperparathyroidism usually manifests clinically before ZES and should be surgically corrected prior to treatment of gastrinoma.

Diagnosis

Three tests are used for the diagnosis of a gastrinoma: fasting serum gastrin concentration, secretin stimulation test, and gastric acid secretion studies. The first two studies are used routinely. A serum gastrin level without a fasting state or stopping antisecretory medications is measured. The upper limit of normal is 110 pg/mL. *A serum gastrin level greater than 1,000 pg/mL is almost diagnostic*. This should be accompanied by a single measurement of gastric pH to ensure that hypergastrinemia is not being caused by achlorhydria. The diagnosis can be confirmed in a patient with a nondiagnostic fasting serum gastrin concentration in whom clinical suspicion of a gastrinoma is high with a secretin stimulation test. The basal serum gastrin is measured and then a 2-unit/kg bolus of secretin is administered. Repeat gastrin levels are obtained at 5-min intervals. An increase in serum gastrin level above 200 pg/mL is considered diagnostic. Serum chromogranin A, a marker for well-differentiated neuroendocrine tumors, is elevated in most patients with gastrinomas and tends to correlate with tumor volume, though it is less sensitive and specific than serum gastrin level.

Localization

Sixty to ninety percent of gastrinomas are located within the gastrinoma triangle—an area between the junction of the cystic duct and common bile duct, the junction of the head and neck of the pancreas, and the second and third

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_27, © Springer New York 2015

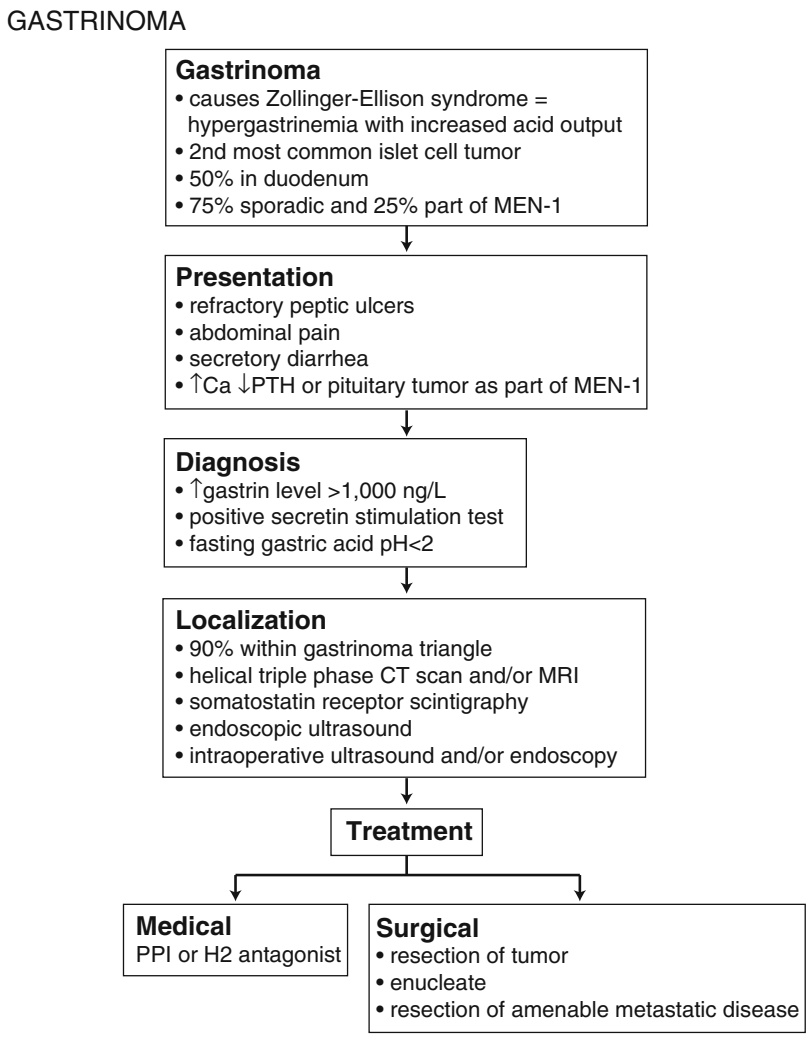

FIG. 27.1 Treatment algorithm for gastrinoma. *MEN-1* multiple endocrine neoplasia type 1, *PTH* parathyroid hormone, *CT* computed tomography, *MRI* magnetic resonance imaging, *PPI* proton pump inhibitor

portion of the duodenum. A gastrinoma and its metastases should be localized preoperatively. Contrast-enhanced triple-phase *computed tomography* (*CT) scan* and *magnetic resonance imaging (MRI)* are used initially to image the pancreas. *Endoscopic ultrasound* can be useful in localizing smaller pancreatic tumors and tumors of the duodenum. In addition, it allows for fine needle aspiration for histological diagnosis. A somatostatin receptor scintigram (using radiolabeled octreotide) has been shown to be superior to CT scan for localizing gastrinomas. About 90 % of gastrinomas have receptors for somatostatin making this a very sensitive test for imaging both primary and metastatic disease. If a strong clinical suspicion for gastrinoma is present and all of the above imaging is negative, other techniques such as angiography and selective arterial stimulation and venous sampling can be performed. If preoperative localization is not possible, intraoperative endoscopy and ultrasound can be used to help localize tumors.

Medical Management

Acid suppression should be initiated as soon as possible in order to prevent complications and to relieve symptoms. Proton pump inhibitors (PPIs) are very effective in suppressing acid production. Oral H2 antagonists can also be used.

Surgical Treatment

The administration of PPIs has made total gastrectomy for gastrinomas almost unnecessary. Patients with a sporadic gastrinoma who do not have evidence of metastatic disease should be offered exploration and resection. Localization prior to surgery is important. Smaller tumors in the pancreas should be treated with enucleation, if possible. Larger tumors in the tail of the pancreas can be treated with a distal pancreatectomy. A pancreaticoduodenectomy is

considered for tumors located in the head of the pancreas; however, it is not without morbidity and mortality itself. Tumors in the duodenal wall should be identified at the time of laparotomy and removed with a duodenotomy and full-thickness excision. In addition, peripancreatic, periduodenal, and portohepatic lymph nodes should be inspected and removed. The most important determinant of survival is the presence of hepatic metastases. Gastric secretion may not return to normal after resection due to the residual excess of parietal cells, and patients may require prolonged antisecretory therapy to control acid secretion following curative resection.

For patients with MEN-1 and gastrinoma, surgery is not generally offered due to the multifocal nature of tumors and the difficulty in resecting all of them with curative intent. These patients are treated medically with acid suppression, and if surgery is undertaken, hyperparathyroidism should be ruled out, and if found, a subtotal parathyroidectomy performed.

Mortality from gastrinomas is related to its malignant nature and the presence of metastases. The treatment of metastatic disease has been of limited benefit. Radiation may be useful for symptomatic palliation for patients who are not surgical candidates. Patients with isolated liver metastases can undergo resection if the number of metastases is limited. Octreotide may be helpful to help reduce gastrin levels and slow tumor growth. Chemotherapy may be of limited benefit and is undertaken on an individualized basis due to the toxicity and uncertainty of its efficacy.

28
Surgical Hypertension: Evaluation and Treatment

Leon Boudourakis and Kaare J. Weber

Background: Signs and Symptoms

Hypertension affects approximately 60 million people in the United States. Long-term sequelae include cardiac arterial disease, congestive heart failure, left ventricular hypertrophy, renal disease, cerebral vascular accidents, and retinopathy. The vast majority of people with hypertension have so-called primary or essential hypertension, whereby no single etiologic cause is known.

When hypertension develops as a result of an identifiable cause, it is termed "secondary" hypertension. This should be suspected in patients who have medically refractory hypertension, an acute rise in blood pressure over a short period of time, are less than 30 years old and without a family history, and those with severe hypertension. Secondary hypertension can be divided into surgical and nonsurgical.

Nonsurgical causes of secondary hypertension include polycythemia vera, hypothyroidism, chronic sleep apnea, as well as renal parenchymal disease such as polycystic kidneys, diabetic nephropathy, glomerulonephritis, and obstructive uropathy. Medications that cause hypertension include corticosteroids, nonsteroidal anti-inflammatory drugs (NSAIDs), estrogens (including several oral contraceptives), cyclosporine, erythropoietin, nasal decongestants, alcohol, amphetamines, ecstasy (MDMA and derivatives), and cocaine.

Workup begins with a thorough history, physical examination, and focused laboratory and radiographic tests (Fig. 28.1).

Renal Renovascular Disease

The majority of patients with renal artery stenosis are men over the age of 45 with atherosclerotic disease involving the aortic orifice or proximal main renal artery. The next most common etiology is fibromuscular dysplasia, which affects women more than men and more often involves the distal main renal artery or intrarenal branches.

As a result of decreased perfusion, juxtaglomerular cells near the glomerulus release renin. This proteolytic enzyme activates angiotensinogen to produce angiotensin I, which is converted to angiotensin II in the lung, ultimately resulting in vasoconstriction and increased adrenal production of aldosterone. Aldosterone in turn causes sodium reabsorption and potassium excretion in the kidney.

Renal artery stenosis can be unilateral (Goldblatt's kidney) or bilateral. The American College of Cardiology and American Heart Association (ACC/AHA) guidelines recommend screening only in those patients in whom an intervention would be seriously considered. This includes patients with hypertension refractory to medications as well as younger patients.

Plasma levels of renin are no longer used as an initial screening tool because of lack of sensitivity. Instead, workup should begin with duplex Doppler ultrasonography, computed tomographic (CT) angiography, or magnetic resonance angiography (MRA) depending on institutional expertise and availability. *Arteriography may be necessary to delineate subtle anomalies and is considered the gold standard.* Stenoses due to fibromuscular dysplasia respond better to angioplasty than those caused by atherosclerosis. Midrenal artery lesions may be treated with angioplasty; ostial lesions, however, do not respond as favorably. Stents may be used for ostial lesions with some success. When these measures fail, bypass or endarterectomy may be beneficial.

Coarctation of the Aorta

Coarctation of the aorta is the narrowing of the medial layer of the vessel. The etiology is most commonly congenital, though it may also be acquired (secondary to Takayasu arteritis or rarely severe atherosclerosis). While this disease can affect neonates, infants, and children, this summary will focus on adults.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_28, © Springer New York 2015

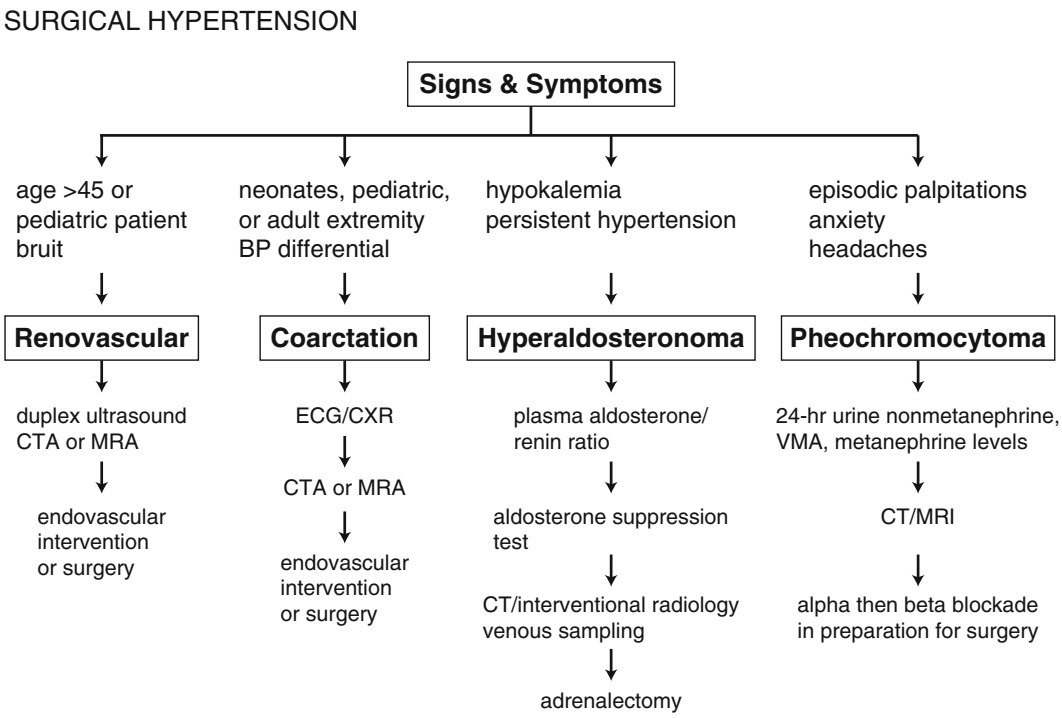

FIG. 28.1 Treatment algorithm for surgical hypertension. *BP* blood pressure, *CTA* computed tomography angiography, *MRA* magnetic resonance angiography, *ECG* electrocardiogram, *CXR* chest X-ray, *VMA* vanillylmandelic acid, *MRI* magnetic resonance imaging

Coarctation most commonly occurs just distal to the left subclavian artery. Physical examination can be significant for blood pressure differences between the left and right arms (proximal coarctation) or between arms and legs (distal coarctation).

Electrocardiographic findings may show left ventricular hypertrophy (increased voltage and ST and T wave changes in left precordial leads). Chest radiograph may reveal a normal or enlarged heart, and there may be notching of the posterior one-third of the third to eighth ribs due to erosions and indentations by tortuous collateral vessels. Transthoracic echocardiography with views from the suprasternal notch can establish the diagnosis, identify significant pressure gradients, and define the location and severity of the coarctation. CT or MRI can also delineate the anatomy.

Definitive management involves percutaneous catheter-based repair (angioplasty and/or stent placement) or surgical repair. According to ACC/AHA guidelines, this is indicated in any patient with imaging evidence of significant coarctation or collateral flow, as well as for patients with peak to peak coarctation gradients above 20 mmHg. The most common surgical repair includes a resection of the coarctation with end-to-end anastomosis. If the length of the coarctation precludes this option, a bypass graft across the area of narrowing can be performed.

Primary Hyperaldosteronism

Primary hyperaldosteronism is the pathologic secretion of aldosterone (independent of renin plasma levels) from unilateral or bilateral adrenal adenomas (Conn's syndrome), bilateral adrenal hyperplasia, adrenal carcinoma, or ectopic aldosterone-producing tumors (neoplasms in ovary or kidney). Secondary hyperaldosteronism occurs due to increased renin levels from congestive heart failure, cirrhosis, ascites, or renin-secreting tumors.

Primary hyperaldosteronism is the most common cause of surgically correctable hypertension and is thought to be underdiagnosed worldwide. It should be suspected in patients with hypertension and a family history of early onset hypertension or primary hyperaldosteronism, concurrent hypokalemia, hypernatremia, or serendipitously discovered adrenal incidentaloma. Patients with drug-resistant hypertension should also be screened.

Clinically, primary hyperaldosteronism should be divided into unilateral or bilateral adrenal disease. Goals of therapy include normalization of blood pressure and hypokalemia. Unilateral disease should be treated surgically, whereas bilateral, benign disease should generally be treated pharmacologically.

The workup of hyperaldosteronism is complex and depends, in part, on institutional expertise and resources and should always be performed in consultation with an experienced endocrinologist. Initial evaluation begins with measuring a plasma aldosterone to renin ratio. If this is elevated (greater than 50), the next step generally involves performing an aldosterone suppression test (orally or intravenously) and measuring urinary excretion or plasma levels.

CT can be used as an initial test to localize adrenal lesions. Bilateral adrenal vein sampling may also be performed. This test relies on the fact that aldosterone secretion is high on the side of an adenoma and is suppressed on the opposite side. Sampling requires an experienced interventional radiology or vascular surgery consultant.

Treatment of choice for unilateral adrenal-producing tumors is laparoscopic adrenalectomy. This can be performed with laparoscopic instruments placed through the peritoneum in the standard fashion or in a retroperitoneal fashion with the patient placed in the prone position, whereby entry into the peritoneum is avoided altogether. The choice depends on surgeon expertise, patient preferences, and the past surgical history of the patient. Standard open laparotomy is also always a viable option.

After adrenalectomy, hypertension is improved in many patients as manifested by decreasing numbers and dosages of antihypertensives. Up to 50–75 % of patients may be cured in 1 year. Hypokalemia is corrected in virtually all patients.

Pheochromocytoma

Pheochromocytoma is a catecholamine-secreting tumor of neural crest origin, which most commonly arises from the adrenal medulla (>90 %). Extra-adrenal sites include the paraaortic region, organ of Zuckerkandl at the bifurcation of the aorta, hilum of the liver or kidneys, urinary bladder, and sympathetic ganglia. Most are benign, though 10 % are malignant. They occur in all ages but are most commonly seen in young to mid-adult life. All patients with pheochromocytoma should be screened for multiple endocrine neoplasia syndrome (MEN) type 2 (constellation of pheochromocytoma, medullary thyroid carcinoma, and hyperparathyroidism) via *RET* proto-oncogene genetic testing.

Secretion of catecholamines (mostly norepinephrine, but also epinephrine and dopamine) may be precipitated by anxiety, trauma, drugs, food, surgery, or anesthesia. Symptoms most commonly include paroxysmal hypertension or chronic hypertension resistant to antihypertensive medications, heart palpitations, headache, and sweating. Other symptoms include visual blurring, weight loss, polyuria, polydipsia, and constipation or diarrhea. Up to 50 % of patients diagnosed with pheochromocytoma are asymptomatic, in which case the lesions are found during workup for an incidentaloma.

Initial workup includes a 24-h measurement of urinary normetanephrines or metanephrines and urinary vanillylmandelic acid (VMA) and plasma catecholamine levels (norepinephrine and epinephrine). Diagnosis is secured if levels are markedly elevated. CT/MRI can localize the tumor. In the setting of biochemically proven disease and high clinical suspicion without CT/MRI evidence of tumor, 123-I-metaiodobenzylguanidine (MIBG) scintigraphy can be used for localization.

Treatment consists of laparoscopic adrenalectomy by an experienced endocrine or laparoscopic surgeon familiar with the disease. Preoperative control of blood pressure is necessary with phenoxybenzamine (alpha-blocking medication). *Beta blockers may be used for tachycardia and hypertension, but only after alpha blockers have been initiated and adequately dosed*. Dexmedetomidine (trade name Precidex), an alpha 2 agonist, is an intravenous sedative with a side-effect profile which may be very beneficial in this patient population pre-, intra-, and post-operatively - these include decreasing sympathetic tone and slowing of the heart rate. Administration needs to be in a monitored setting. Calcium channel blockers can also be used in place of alpha blockers in select clinical scenarios. Failure to do so may result in precipitating a hypertensive crisis if not all alpha receptors are blocked.

Patients with MEN 2 and evidence of bilateral disease on imaging should undergo complete bilateral adrenalectomy. Patients with von Hippel-Lindau (VHL) with bilateral disease should undergo cortical-sparing bilateral adrenalectomy (unless there is a high risk of malignancy).

Miscellaneous Surgical Diseases that Contribute Hypertension

Patients with hypertension and concurrent hyperthyroidism, hyperparathyroidism, and adrenocorticotropic hormone (ACTH)-secreting tumors (including pituitary adenomas and adrenocortical adenomas/carcinomas) may have their hypertension lessened after surgical cure. It should be noted, however, that most of these patients have primary hypertension with these secondary disease processes contributing to their level of systolic/diastolic derangement.

Tumors secreting corticotropin-releasing hormone (CRH) by nonhypothalamic tumors cause secondary pituitary hypersecretion of ACTH.

29
Incidentalomas

John Cull and Katherine Heiden

Introduction

An adrenal incidentaloma is an adrenal mass larger than 1 cm discovered on an imaging study obtained for purposes other than evaluating the adrenal glands. With the increasing use of computed tomography (CT) and magnetic resonance imaging (MRI), the detection of clinically occult adrenal masses has increased over the past 20 years. In the general population, approximately 1 % of healthy patients will have an adrenal mass. The incidence of adrenal masses increases with age to approximately 10 % in the elderly. In patients with a prior history of malignancy, approximately 4 % will have an adrenal mass. The goal in the workup of adrenal masses is to identify those masses that are functional or likely to be malignant, to remove them, and to observe the rest (Fig. 29.1). *To determine if an adrenal mass is functional, all patients with adrenal masses should be screened for hyperaldosteronism, hypercortisolism, and pheochromocytoma.*

Hyperaldosteronism

Hyperaldosteronism is caused by an overproduction of aldosterone from the zona glomerulosa of the adrenal glands. When it is due to an adrenal adenoma, it is referred to as Conn's syndrome. Aldosterone increases the number of sodium-potassium pumps in the distal tubules and collecting ducts of the nephron, thereby resulting in sodium retention and excretion of potassium. Patients may present with muscle weakness and fatigue due to hypokalemia. Patients can also present with diastolic hypertension, polydipsia, and polyuria. *Hyperaldosteronism should be considered in patients with hypertension and hypokalemia.* A hallmark of hyperaldosteronism is hypertension without peripheral edema. Hyperaldosteronism is diagnosed by obtaining a plasma aldosterone to renin ratio; a value greater than 30 is diagnostic.

Hypercortisolism

Hypercortisolism is caused by an overproduction of cortisol from the zona fasciculate of the adrenal glands. Cushing's syndrome refers to hypercortisolism from any cause (medications, pituitary adenomas, cancers) including cortisol-producing adrenal adenomas. Cushing's disease refers specifically to hypercortisolism that is due to a pituitary adenoma. The signs and symptoms of Cushing's syndrome or hypercortisolism include truncal obesity, moon facies and buffalo hump, muscle wasting, hirsutism, hypertension, osteoporosis, headache, emotional liability, depression, and psychosis. Hypercortisolism can be diagnosed in several ways. A 24-h cortisol urine result greater than 100 μg is diagnostic. A low-dose dexamethasone test can also be used for diagnosis. One milligram of dexamethasone is administered orally at 11 PM and a plasma cortisol is obtained at 8:00 AM the following day. If the plasma cortisol is above 5 μg/dL, the patient has Cushing's syndrome. Patients with Cushing's syndrome should receive glucocorticoid therapy perioperatively to reduce the risk of adrenal insufficiency.

Pheochromocytoma

A pheochromocytoma secretes an excessive amount of catecholamines (norepinephrine and epinephrine) from the medulla of the adrenal gland. Pheochromocytomas are present in familial diseases such as MEN2A, MEN2B, von Hippel-Lindau, and neurofibromatosis type 1. Pheochromocytomas are described as following the "rule of 10s": 10 % of pheochromocytomas are bilateral, 10 % are extrarenal, 10 % familial, 10 % malignant, and 10 % occur in children.

The classic triad of symptoms is episodic headaches, sweating, and tachycardia. Patients may have hypertension. Patients are generally hypovolemic, and orthostatic hypotension may result from decreased plasma volumes. Patients

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_29, © Springer New York 2015

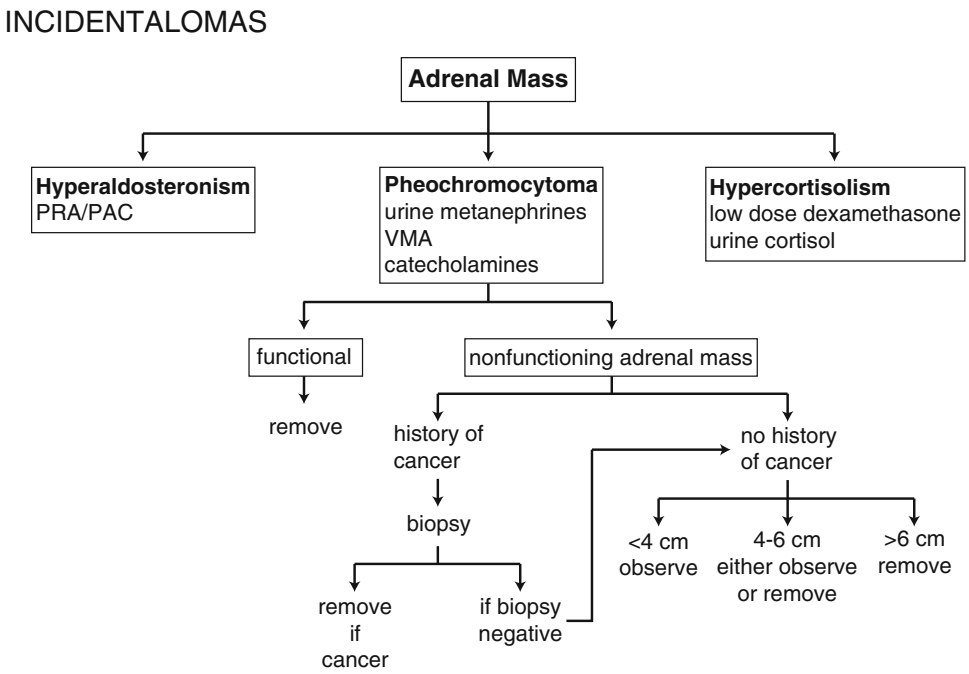

FIG. 29.1 Treatment algorithm for adrenal mass. *PRA* plasma renin activity, *PAC* plasma aldosterone concentration, *VMA* vanillylmandelic acid

may also experience palpitations, anxiety, cardiac arrhythmias, and stroke. Sudden cardiac death has been reported in patients with pheochromocytoma who have undergone surgical procedures or childbirth.

Features on CT that suggest a pheochromocytoma are increased attenuation on unenhanced CT, prominent vascularity of the mass, and delayed washout of contrast. Pheochromocytomas will also have a high signal intensity on T2-weighted MRI. The diagnosis is established with 24-h urine tests for catecholamines, metanephrines, and VMA. Plasma metanephrines can also be obtained in cases when the suspicion of a subclinical pheochromocytoma is high based upon imaging despite normal urine catecholamines. Plasma metanephrines have a high sensitivity for detecting pheochromocytoma (96–100 %), but the test has a low specificity (85–89 % overall and 77 % in patients older than 60 years). It is important not to biopsy pheochromocytomas because this may lead to complications arising from a potentially massive release of catecholamines.

Prior to surgical resection for pheochromocytomas, patients are placed on alpha-adrenergic blockade with phenoxybenzamine 1–3 weeks prior to operation. Alpha blockade helps reverse hypovolemia and prevents severe pressure swings intraoperatively. Beta blockade with propranolol can be added after alpha blockade to help manage patients who develop tachycardia or who have cardiac arrhythmias/ischemia.

After ruling out a functional adrenal adenoma, the adrenal mass should be evaluated for possible cancer. The adrenal glands are frequent sites of metastases from lung/breast cancer but may also occur from melanoma, lymphoma, kidney, and ovarian cancer. Among cancer patients, 50–75 % of newly diagnosed adrenal masses are found to be metastases. In patients with a history of carcinoma and an adrenal mass, a biopsy of the adrenal mass should be performed to rule out metastasis *after a pheochromocytoma is excluded*.

Excision is indicated for all functional adrenal incidentalomas and those masses suspected of being cancerous. Lesions larger than 6 cm are cancer in approximately 25 % of patients and should be resected. Adrenal masses less than 4 cm have a less than 2 % chance of being cancerous and can be observed. Lesions between 4 and 6 cm can either be resected or observed based upon the clinical situation. For instance, in an 80-year-old patient, a nonfunctioning 5 cm adrenal mass can be reasonably followed, while a 4 cm mass in a 25-year-old patient should have a very low threshold for removal.

Conclusion

A through history and physical examination should be performed on all patients with an incidentaloma to identify evidence of a functional adrenal adenoma. A plasma aldosterone to renin ratio should be measured to rule out hyperaldosteronism.

A 24-h urine cortisol test or an overnight dexamethasone test should be obtained to rule out hypercortisolism, and urine metanephrines and catecholamines should be measured to rule out pheochromocytoma. In patients with a history of carcinoma, a CT-guided biopsy should be considered after a pheochromocytoma is ruled out. *Nonfunctioning adrenal masses less than 4 cm can be observed while masses greater than 6 cm should be removed. Adrenal masses between 4 and 6 cm can be either resected or observed based upon the clinical situation*.

Part V
Abdomen

30
Upper Gastrointestinal Hemorrhage

Nikolaos D. Karagiorgos and Matthew J. Hyser

Introduction

Upper gastrointestinal hemorrhage has a variety of causes and may present with subtle or massive blood loss. Potential sources may be located anywhere from the nasal/oropharynx to the ligament of Treitz. The history and physical examination are cornerstones for initial assessment; if the patient's vital signs are unstable, resuscitation and evaluation are promptly instituted simultaneously with a rapid physical assessment and interview with the patient and/or family (Fig. 30.1). A variety of over-the-counter or prescribed medicines may either cause GI bleeding or lead to failure of hemostasis during treatment. A history of their use is essential.

Intravenous crystalloid resuscitation is begun through two large-bore peripheral catheters (16 gauge). Blood for hemoglobin, platelet count, PT, PTT, and type and cross is obtained. Simultaneously, the patient's airway is assessed and an 18-gauge nasogastric (NG) tube and bladder catheter are placed.

Significant hematemesis, tachycardia, or hypotension should prompt early intubation to secure the airway and prevent aspiration. Room temperature saline gastric lavage is performed, and the response to initial management is evaluated.

Patients with massive bleeding and hemodynamic instability should be intubated and transferred to the operating room. Further resuscitation and esophagogastroduodenoscopy (EGD) can be rapidly performed there before exploratory laparotomy. If endoscopy reveals esophageal varices as the source of hemorrhage, laparotomy may be withheld in favor of an attempt at either endoscopic variceal band ligation (EVL), endoscopic sclerotherapy (EST), tamponade with a Sengstaken-Blakemore tube, or transjugular intrahepatic portosystemic shunt (TIPS). Critical points in the use of a Sengstaken-Blakemore tube include endotracheal intubation to protect the airway, adequate inflation of the gastric balloon in the stomach to avoid tube migration, and avoidance of esophageal compromise by monitoring pressure within the esophageal balloon.

Generally accepted indications for surgery include ongoing hemorrhage with instability, 6 units transfusion of packed RBCs during the first 24 h, and failed attempts at endoscopic control. If the decision is made to proceed with surgery for the unstable patient, an upper midline incision is used to enter the abdomen and explore for clues as to the etiology of bleeding. If none are discovered, the stomach is opened longitudinally and a complete inspection is undertaken for ulcers, Mallory-Weiss tears, neoplasms, arteriovenous malformations, or other pathology.

Most patients with upper GI bleeding are stable enough to undergo semielective endoscopy in either the emergency room or the endoscopy suite. An Ewald tube placed into the stomach just prior to endoscopy will efficiently remove blood clots with irrigation and suction, thereby improving visualization. Many sources of hemorrhage can be successfully diagnosed and treated in this fashion. Subsequent admission to an intensive care unit is prudent for monitoring.

Esophageal Varices

Endoscopy with rubber band ligation (preferred over EST) effectively controls hemorrhage from esophageal varices. Repeated sessions may be required. Other therapeutic options include octreotide, beta blockade, and a Sengstaken-Blakemore tube. Consider TIPS if initial therapy fails. Isolated gastric varices may be caused by portal hypertension, portal vein thrombosis, or splenic vein thrombosis (sinistral portal hypertension). Splenectomy is curative for the latter while EVL and EST can be used for acute bleeding in the former. Prevention of infectious complications reduces mortality in cirrhotic patients with upper GI bleeding. Use either norfloxacin 400 mg PO BID for 7 days or ceftriaxone 1 g IV daily.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_30, © Springer New York 2015

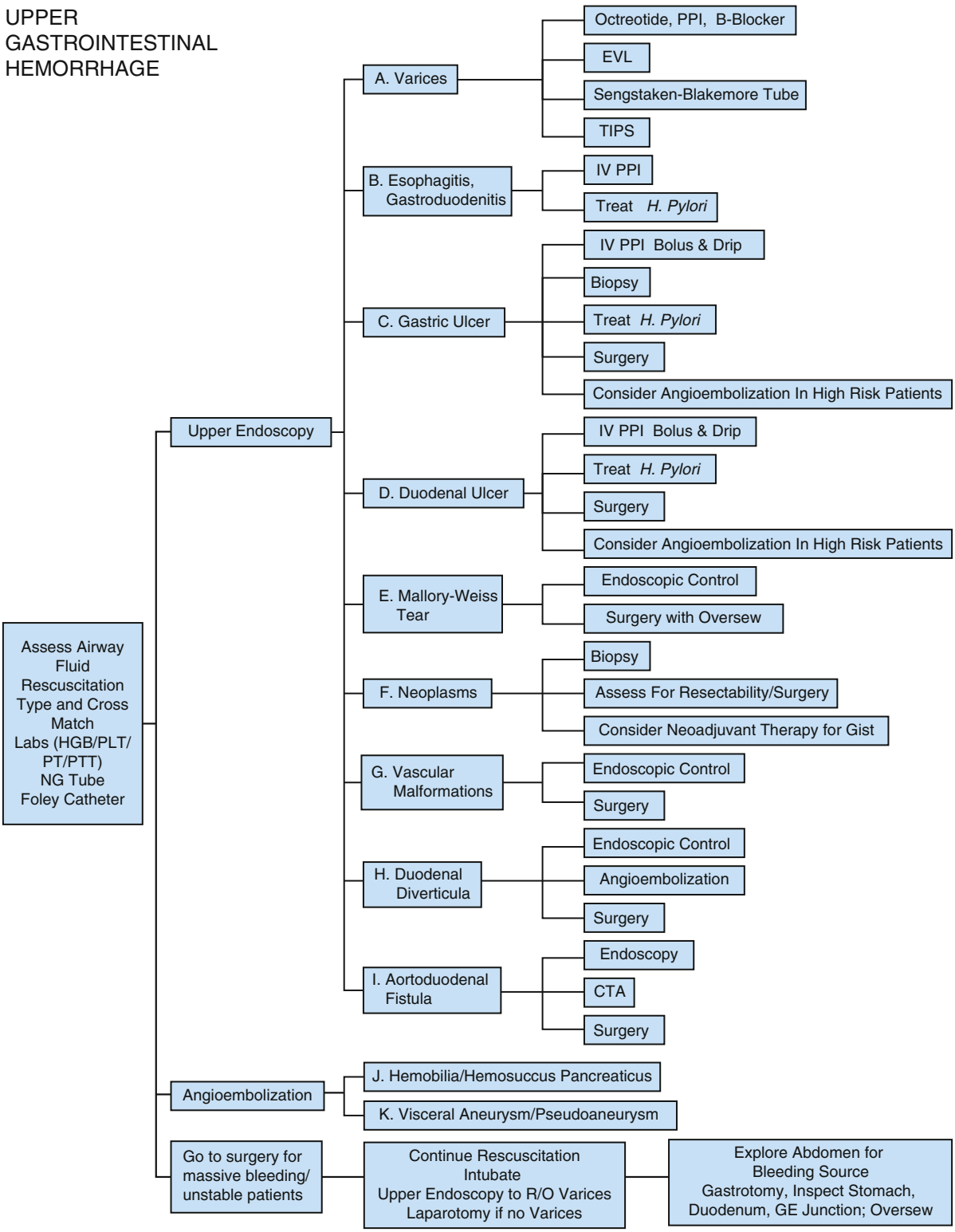

FIG. 30.1 Algorithm for treatment of upper gastrointestinal hemorrhage. *NG* nasogastric, *PPI* proton pump inhibitors, *EVL* endoscopic variceal band ligation, *TIPS* transjugular intrahepatic portosystemic shunt, *IV* intravenous, *CTA* computed tomography angiography

Esophagitis, Gastritis, and Duodenitis

These conditions are treated with oral or intravenous proton pump inhibitors (PPI) together with antacid therapy either by mouth or by NG tube. Tissue biopsy and CLO testing for *Helicobacter pylori* should be performed and antibiotics added if positive. Additionally, any offending medications are discontinued. Diffuse uncontrolled bleeding from stress gastritis requiring operation is now rare. If surgery becomes necessary, the stomach is opened and bleeding points are controlled with suture ligature and Gelfoam with thrombin and/or Surgicel. Truncal vagotomy will decrease mucosal blood flow and should be considered if these maneuvers fail. Partial or total gastrectomy may be required depending upon the predominant sites of bleeding, although this is rarely required.

Gastric Ulcer

Bleeding gastric ulcers are treated with intravenous PPI therapy (pantoprazole 80 mg IV bolus followed by continuous infusion of 8 mg/h for 72 h). Oral antacids can also be used. The endoscopic diagnosis of gastric ulcer is usually straightforward. *It is important to biopsy to rule out malignancy and to diagnose H. pylori infection*. Endoscopic therapies to control bleeding include heater probe, unipolar or bipolar cautery, laser therapy, argon plasma coagulation, epinephrine injection (1:10,000), and endoscopic clip placement. A gastroduodenal ulcer with a visible vessel at its base has a 40–50 % rebleeding rate. A decision to either rescope or operate for subsequent hemorrhage after endoscopy is made at the conclusion of the initial endoscopic session. Rebleeding during the same hospitalization or a transfusion requirement of more than 6 units of red blood cells are considered indications for surgery. Surgical options for bleeding gastric ulcers include oversew, ulcer excision, and hemigastrectomy. Vagotomy is not required but is essential in the treatment of duodenal and prepyloric gastric ulcers. Angioembolization is an alternative for high-risk patients or rebleeding occurring after operative management.

Duodenal Ulcer

The medical and endoscopic treatment of duodenal ulcer is similar to gastric ulcer management: IV PPI bolus followed by continuous infusion for 72 h. *H. pylori*, if detected, is treated. Inability to endoscopically control bleeding or evidence of persistent or recurrent hemorrhage indicates the need for operation. Surgical treatment for bleeding duodenal ulcers includes dividing the pylorus longitudinally with prompt suture control of the bleeding vessel and/or gastroduodenal artery. Vagotomy and pyloroplasty complete the procedure. A large pyloroduodenal ulcer may require antrectomy with vagotomy. As with gastric ulcers, bleeding duodenal ulcers in either high-risk individuals or in those who rebleed postoperatively may be considered for angioembolization.

Mallory-Weiss Tear

A Mallory-Weiss tear occurs at the distal esophagus and extends to the gastric cardia. Often a history of intense vomiting precedes brisk arterial bleeding. Surgery is necessary if endoscopic control fails and consists of high gastrotomy and oversew of the bleeding point.

Neoplasms

A variety of benign and malignant neoplasms of the esophagus, stomach, and duodenum may bleed. Bleeding is usually not torrential at presentation; lesions can be identified, biopsied, and electively resected after evaluation and staging is completed. Gastric leiomyomas or GIST tumors are the most common benign and malignant neoplasms, which may bleed massively and require emergent resection. There is an emerging role for neoadjuvant (preoperative) therapy with Gleevec for GIST tumors in difficult locations.

Vascular Malformations

A variety of vascular malformations may occur in the gastric mucosa. These include arteriovenous malformations, radiation-induced telangiectasia, congestive gastropathy (portal hypertension), antral ectasia (watermelon stomach), and Dieulafoy's arterial malformation. This entity is very hard to recognize if not actively bleeding since there is no associated mucosal lesion present. All may present with upper tract bleeding, are relatively uncommon, and can be controlled with endoscopic hemostatic techniques. Surgery is reserved for treatment failure.

Duodenal Diverticula

Bleeding duodenal diverticula present a special challenge in both diagnosis and therapy. They may be difficult to visualize on routine upper endoscopy and can be found in each portion of the duodenum but commonly arise adjacent to the ampulla of Vater. If endoscopic techniques fail to control hemorrhage, surgery with either direct suture control or diverticulectomy may be required. It is essential to precisely identify the course of the pancreatic and biliary ducts to avoid their injury during surgery. Angioembolization is another therapeutic option.

Aortoduodenal Fistula

Patients with prior aortic graft surgery may present with upper or lower GI hemorrhage, usually as a herald bleed followed by a more significant bleed. This history should prompt immediate evaluation for aortoduodenal fistula. Unstable patients should be taken promptly to the operating room. Stable patients are evaluated with endoscopy and CT angiography (CTA). Chronic infection caused by erosion of graft material into the duodenum may also be identified by tagged WBC nuclear medicine scanning. Pseudoaneurysm formation at the proximal anastomosis or visible graft in the third to fourth portion of the duodenum requires emergency surgery and extra-anatomic arterial reconstruction.

Hemobilia and Hemosuccus Pancreaticus

Unlocalized upper GI bleeding, blood in the duodenum without an apparent source, or blood from the ampulla of Vater may be due to hemobilia or hemosuccus pancreaticus. Liver trauma and indwelling stents may cause the former while inflammatory pseudocysts with erosion and pseudoaneurysm formation cause the latter. Angiography with embolization is diagnostic and therapeutic for both conditions.

Visceral Arterial Aneurysms and Pseudoaneurysms

These entities may produce intestinal, intraperitoneal, or retroperitoneal hemorrhage. Splenic artery aneurysms comprise approximately 60 %, hepatic artery aneurysms approximately 20 %, while celiac, gastric, gastroepiploic, jejunoileal, and gastroduodenal/pancreaticoduodenal arteries produce the remainder of aneurysmal bleeding complications. With the exception of splenic artery aneurysms, all others may present with GI tract hemorrhage. Early evaluation may begin with endoscopy of the GI tract, but CT angiography is the diagnostic study of choice with embolization as first-line therapy. Surgery should follow failed attempts at embolization but may also be required initially for patients presenting with shock with or without a firm preoperative diagnosis.

31
Dysphagia

Michael S. Vercillo and Edward Hong

History and Physical Exam

Dysphagia, or difficulty swallowing, can be very distressing to a patient. A thorough history and physical examination must be done to determine the cause and severity such as difficulty with solids, liquids, or both (Fig. 31.1). One should determine if there is pain with swallowing (odynophagia), emesis of undigested food, history of heartburn or gastroesophageal reflux disease (GERD), history of systemic disease such as scleroderma, and history of caustic ingestion. A differential diagnosis is provided in Table 31.1.

Barium Swallow

A barium swallow study should be the first diagnostic test. Many physicians do not proceed initially with an upper endoscopy until the esophageal anatomy has been delineated through radiographic means. The barium swallow may reveal a primary motility disorder, evidence of esophageal diverticulum, obstruction, or may be normal.

Primary Motility Disorders

Primary motility disorders may be suggested on barium swallow; confirmation is with esophageal manometry and endoscopy. *Achalasia* is caused by a failure of the lower esophageal sphincter (LES) to relax. The classic triad of presenting symptoms includes dysphagia, regurgitation, and weight loss. A typical barium esophagram demonstrates a bird-beak tapering of the distal esophagus associated with varying degrees of proximal dilation.

The second diagnostic procedure is endoscopy to rule out carcinoma. If a mechanical obstruction is not found, esophageal manometry is done. The typical manometric findings include failure of the LES to relax with swallowing and deficient progressive peristalsis within the esophagus, which is present with normal swallowing. *Forceful balloon dilatation* may be used to treat the symptoms of early disease; however, only 65 % of patients will experience long-term relief. If dilatation fails, the treatment of choice is *distal esophageal myotomy combined with an anti-reflux procedure*. The myotomy can be performed through a left thoracotomy or thoracoscopy, transabdominally or laparoscopically (Heller operation). A successful myotomy will divide both longitudinal and circular muscles of the esophagus and is extended through the LES onto the stomach about 2 cm. The incidence of GERD after myotomy can be as high as 50 %; therefore, the myotomy should be accompanied by an anti-reflux procedure (usually a partial fundoplication, i.e., Toupet or Dor). Endoscopic means of performing the myotomy are now being explored (peroral endoscopic myotomy [POEM]).

Diffuse esophageal spasm typically presents with dysphagia and chest pain. Barium esophagram may show a corkscrew esophagus, and manometry shows multi-peaked, high amplitude contractions of long duration. Treatment is medical with calcium channel blockers, nitrates, or anticholinergics. If medical therapy fails, surgical treatment is with a long esophageal myotomy with a partial fundoplication via a left thoracotomy or video-assisted thoracoscopy.

Nutcracker esophagus also presents with dysphagia and chest pain. Patients may also present with odynophagia. Manometry shows frequent contractions at least two standard deviations above normal. Treatment is medical with calcium channel blockers, nitrates, or antispasmodics.

Hypertensive LES is characterized by elevated LES pressure with normal relaxation. It is first treated with endoscopic Botox injections or dilation. If these fail, a myotomy is indicated.

Diverticula

Esophageal diverticula are categorized as either pulsion or traction diverticula. Pulsion diverticula occurring in the upper third of the esophagus are called "Zenker's diverticula."

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_31, © Springer New York 2015

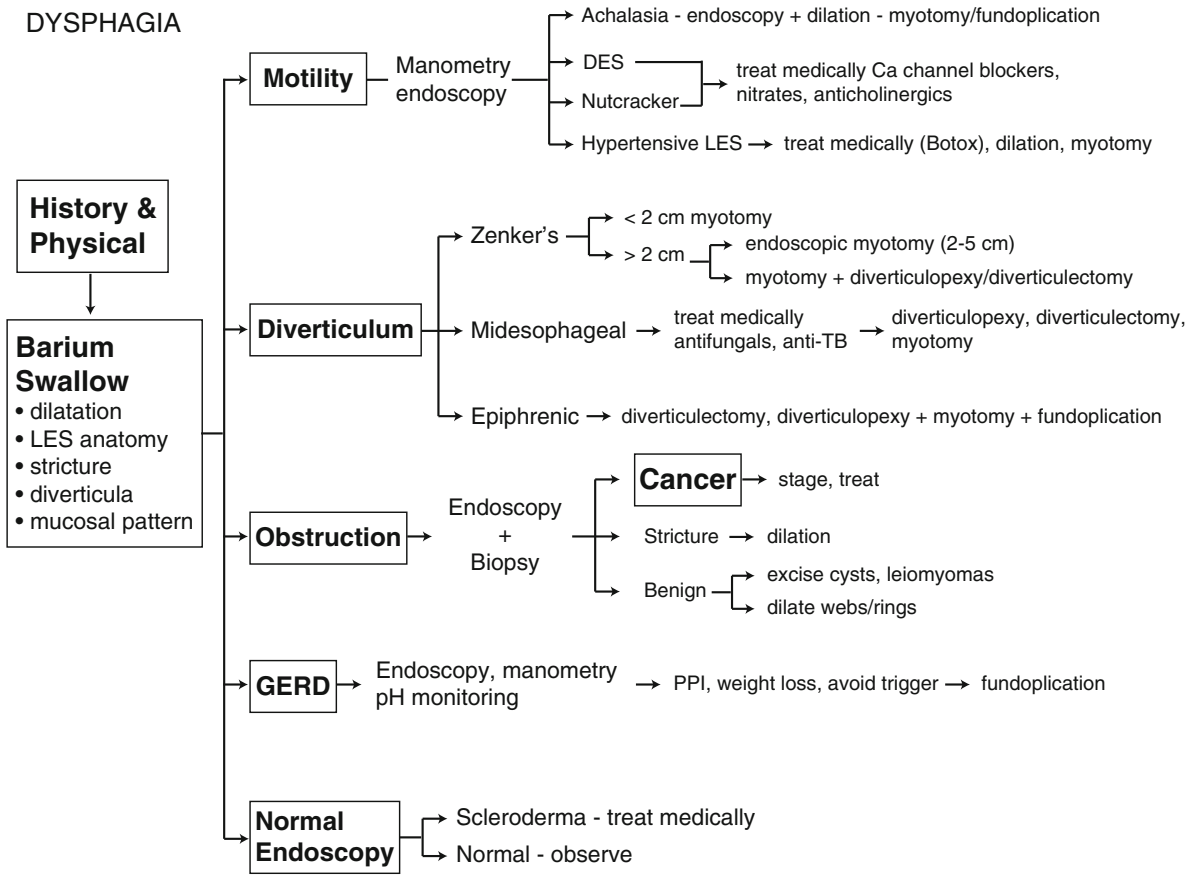

FIG. 31.1 Treatment algorithm for dysphagia. *LES* lower esophageal sphincter, *GERD* gastroesophageal reflux disease, *DES* diffuse esophageal spasm, *TB* tuberculosis, *PPI* proton pump inhibitors

TABLE 31.1 Differential diagnosis of dysphagia

I.	*Anatomic*
	Leiomyoma
	Esophageal web or ring
	Peptic stricture
	Carcinoma
II.	*Physiologic*
	Zenker's diverticulum
	Esophageal traction diverticulum
	Epiphrenic diverticula
	Achalasia
	Diffuse esophageal spasm
	Scleroderma
III.	*Neurologic*
	Chagas disease
	Central deficit/pharyngeal dysphagia

These are false diverticula caused by uncoordinated swallowing that form at a weak point between the longitudinal cricopharyngeus and oblique thyropharyngeus muscles called *Killian's triangle*. Treatment involves cricopharyngeal myotomy and may also include a diverticulectomy or diverticulopexy. Diverticula smaller than 2 cm can be treated with myotomy alone. The diverticulum is approached through a left neck incision and is found posterior to the esophagus. Endoscopic myotomies are being performed more frequently and are ideal for diverticula between 2 and 5 cm. Midesophageal diverticula are wide-mouthed, true diverticula caused by traction on the esophageal wall from inflamed paraesophageal lymph nodes secondary to a process such as histoplasmosis, toxoplasmosis, or tuberculosis. Treatment is aimed at the underlying condition with antifungal or antituberculosis agents. Epiphrenic diverticula occur in the distal third of the esophagus, usually within 10 cm of the LES. They are typically wide-mouthed, false diverticula and are associated with an underlying motility disorder. Treatment involves a diverticulectomy or diverticulopexy with a myotomy and partial fundoplication.

Obstruction

If the barium swallow reveals an obstructing lesion or stenosis, endoscopy with biopsy is indicated. If the lesion is benign and the obstruction is thought to be a peptic stricture from reflux or a stricture from previous ingestion of a caustic solution, dilatation should be performed. Resection is

needed only when the lesion is refractory to dilatation; the colon and the stomach have both been used as replacements for the esophagus. If the biopsy reveals a benign cyst or leiomyoma, excision should be carried out through a thoracic approach. Esophageal webs and rings are treated with endoscopic dilatation or ablative therapy; excision is reserved for those instances when conservative measures have failed. Paraesophageal hernias causing obstruction should be repaired with a reduction of hernia and fundoplication.

Cancer

If cancer is diagnosed, a metastatic survey including computerized tomography (CT) of the neck, chest, and abdomen should be performed. Endoscopic ultrasound (EUS) may assess depth of tumor invasion and for periesophageal or celiac axis lymph node metastases. A positron emission tomography (PET) scan may also aid in staging. Patients with early stage cancer (T1N0) may be treated with esophagectomy alone. More advanced lesions or those with local lymph node involvement are treated with neoadjuvant chemotherapy and radiation followed by resection. Distant metastasis or invasion into adjacent structures is considered unresectable disease. Many patients present with advanced disease, and the treatment in these cases is aimed at palliation—options include endoscopic esophageal stenting or laser ablative therapy in addition to chemotherapy and radiation.

Gastroesophageal Reflux

Patients with GERD may present with dysphagia. Following endoscopy and manometry, pH monitoring is the next diagnostic test. Medical treatment is begun first with proton pump inhibitors, weight loss, and avoidance of aggravating factors such as caffeine, alcohol, and tobacco. Patients with continued symptoms may be treated with a partial or complete fundoplication.

32
Esophageal Cancer

Rana M. Ballo and Keith W. Millikan

Epidemiology

In 2011, approximately 17,000 new cases of esophageal cancer were diagnosed in the United States and 15,000 died from this disease. Five-year survival is estimated at 17 %, increased from 5 % in the 1970s. The average age of diagnosis is 68. The two major histologic types are *squamous cell carcinoma* (SCC) and *adenocarcinoma*. The highest incidence rates are found in the "esophageal cancer belt" of Asia, Africa, and Iran. In these areas, 90 % of cases are squamous cell carcinoma, whereas in the United States, adenocarcinoma is more prevalent. Men are eight times more often affected than women. Risk factors for squamous cell carcinoma include smoking, alcohol abuse, foods containing *N*-nitroso compounds, drinking hot tea, achalasia, tylosis, and a history of caustic esophageal injury in childhood. The predominant risk factor for adenocarcinoma is Barrett's esophagus (30-fold increased risk), which is intestinal metaplasia that occurs as a result of long-standing gastroesophageal reflux disease (GERD).

Presentation

Esophageal cancers are aggressive with early local invasion to lymphatics. Patients present primarily with symptoms of dysphagia and weight loss (Fig. 32.1). Additional symptoms include retrosternal pain, regurgitation of saliva or food, chronic blood loss leading to iron deficiency anemia, and tracheobronchial fistulas leading to frequent coughing and pneumonias. It is estimated that 50–60 % of patients with esophageal cancer present with locally unresectable or metastatic disease, for which palliation would be the primary focus.

Diagnosis

Barium swallow delineates the anatomy, but endoscopy is needed to confirm the diagnosis with biopsy or brush cytology. The primary site of SCC is the middle esophagus, whereas the distal third of the esophagus and GE junction are the most common sites for adenocarcinoma. Once a tissue diagnosis is obtained, locoregional staging is best performed with endoscopic ultrasound (EUS), which is helpful in determining depth of invasion (T stage) as well as lymph node involvement (N stage). If malignant-appearing lymph nodes are identified, fine needle aspiration (FNA) can be performed for cytologic analysis. Metastatic disease is assessed with computed tomography (CT) and/or positron-emission tomography (PET) scans evaluating the neck, chest, and abdomen. PET scans have greater sensitivity for detecting metastatic disease; the most common locations are the liver, lungs, bone, and adrenal glands. Additionally, because of proximity to the trachea and bronchus, all patients with thoracic lesions should undergo preoperative bronchoscopy.

Staging

See Table 32.1 for tumor node metastasis (TNM) staging.

Treatment

A multimodal approach is often used. Localized, intramucosal tumors can be treated with endoscopic mucosal resection (EMR). Esophagectomy is also an option, but its high morbidity makes EMR a more favorable option. As long as there is no invasion into the muscularis mucosa, EMR is a definitive treatment. These patients, however, will require lifetime annual surveillance with endoscopy.

For ultrasound T2 lesions, a debate continues between prompt esophagectomy versus neoadjuvant chemoradiation followed by surgery. For younger, healthier patients, a more aggressive approach with neoadjuvant therapy is considered to see if a complete response could be obtained, as this has demonstrated better long-term survival. Older, more debilitated patients may not tolerate neoadjuvant therapy; therefore, immediate surgical resection is performed; adjuvant

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_32, © Springer New York 2015

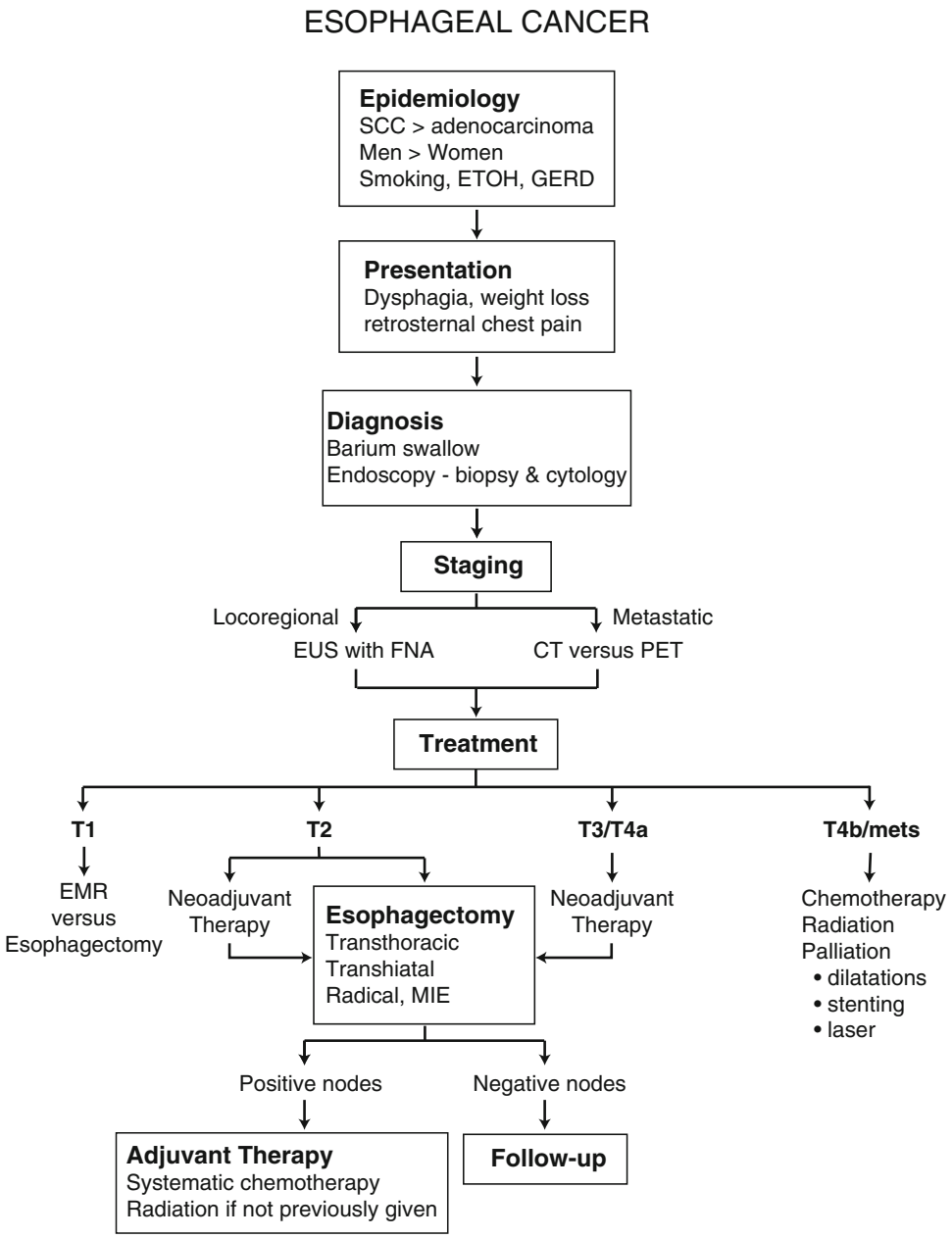

FIG. 32.1 Esophageal cancer algorithm. *SCC* squamous cell carcinoma, *GERD* gastroesophageal reflux disease, *EUS* endoscopic ultrasound, *FNA* fine needle aspiration, *CT* computed tomography, *PET* positron-emission tomography, *T* tumor stage, *EMR* endoscopic mucosal resection, *MIE* minimally invasive esophagectomy

TABLE 32.1. 2010 TNM staging for esophageal cancer.

Invasion of primary tumor (T)	Lymph nodes (N)	Metastases (M)
T1: lamina propria, muscularis mucosa, or submucosa	N1: 1–2	M1: distant metastases
T2: muscularis propria	N2: 3–6	
T3: adventitia	N3: 7 or greater	
T4: adjacent structures (T4a: resectable – pleura, pericardium, or diaphragm T4b: unresectable – aorta, vertebral body, trachea, etc.)		

therapy is indicated for patients who are found to have T3 or higher disease or lymph node metastases.

For T3 and T4a lesions, neoadjuvant chemoradiation is pursued followed by surgical resection. Patients with metastatic disease on presentation or node-positive disease after surgery are candidates for chemoradiation therapy, in addition to palliative measures, such as esophageal dilation or stenting. The combination of chemotherapy and radiation therapy has shown improved survival compared to either alone.

Surgical Resection

There are several options for surgical resection of esophageal cancer. Cervical tumors are primarily SCC and are evaluated for tracheal and laryngeal involvement prior to primary resection. Additionally, given the difficulty in obtaining negative margins, most of these patients are given neoadjuvant therapy prior to surgical resection. The management of these tumors is similar to head and neck SCCs. Thoracic tumors are resected via transthoracic or transhiatal approaches.

Transthoracic esophagectomy (Ivor-Lewis) was the first type of resection performed for esophageal cancer. This approach requires two incisions: laparotomy and right thoracotomy. Initially, via a laparotomy incision, the stomach and intra-abdominal esophagus are mobilized. In the second stage, a right thoracotomy is performed and the esophagus is mobilized, with the creation of an intrathoracic esophagogastric anastomosis. The primary tubularized conduit for esophageal reconstruction is usually gastric, although jejunal and colonic interpositions are alternate possibilities. The morbidities of this procedure are primarily pulmonary, including pneumonia, pleural effusions, and intrathoracic anastomotic leak. Intrathoracic anastomotic leak has a high risk of sepsis and is often a challenge to manage successfully. For midthoracic tumors, transthoracic is the preferred approach because unrecognized vascular and tracheobronchial involvement is best identified via this approach.

An alternate resection is *transhiatal esophagectomy* (THE) and is frequently utilized for distal third tumors. The approach requires two incisions: laparotomy and left neck. The intrathoracic esophagus is mobilized, and a cervical esophagogastric anastomosis is performed. The advantages of this procedure are fewer respiratory complications and an anastomotic leak, if it should occur, that is more easily managed because of its cervical location.

A third, more aggressive approach described is *radical esophagectomy*, which requires three incisions: left neck, right chest, and abdomen. Radical lymphadenectomy is an added component. This approach seems to be favored in Japan where increased survival data have been noted. This has not been seen in Western countries. Minimally invasive esophagectomy (MIE) has been gaining popularity over the last several years, using both thoracoscopic and laparoscopic approaches. Unresectable esophageal cancers can be palliated with chemoradiation, stenting, or laser therapy provided via a gastrostomy or jejunostomy tube.

Follow-Up

After esophagectomy, patients are followed with surveillance CT or PET scans every 6 months for the first 2 years and then annually. Additionally, routine labs are obtained, including CBC, liver function panels, and CEA. Endoscopy may be performed as well if symptoms of dysphagia occur.

33
Gastroesophageal Reflux

Andrew M. Popoff and Daniel J. Deziel

Introduction

Gastroesophageal reflux disease (GERD) is a common medical problem routinely encountered by physicians across the continuum of care. Its incidence is estimated to be between 10 and 22 % in Western populations, with that number increasing to 40 % when patients with mild symptoms are included. Typical symptoms of GERD include heartburn, regurgitation, dysphagia, chest pain, and water brash (hypersalivation). Extraesophageal manifestations of GERD include odynophagia, chronic cough, asthma, laryngitis, globus hystericus, or dental erosions. With the widespread adoption of minimally invasive surgical techniques, surgery is playing an increasingly important role in the management of GERD.

Patient Presentation, Evaluation, and Initial Management

Patients presenting with heartburn and regurgitation are commonly and acceptably treated empirically (Fig. 33.1). For mild, intermittent symptoms, over-the-counter antacids and H2 blockers may be sufficient. For more regularly occurring symptoms, empiric therapy with a PPI for 6–8 weeks is reasonable. Response to therapy, while not diagnostic, is highly presumptive. Patients are counseled regarding lifestyle modifications including weight loss, diet alteration, and avoidance of precipitating foods and beverages, such as caffeine, nicotine, alcohol, and fat. Patients are instructed to refrain from oral intake 2–3 h before bedtime and to elevate the head of the bed 6–10 in.

Significant dysphagia, weight loss, or anemia suggests complicated GERD with esophageal stricture or an alternative diagnosis, such as an esophageal motility disorder or neoplasm. Patients with these findings warrant diagnostic investigation with esophagogastroduodenoscopy (EGD), radiologic contrast study of the esophagus and upper gastrointestinal tract, and esophageal manometry. Esophageal pH monitoring can confirm or exclude abnormal acid exposure. Patients with symptoms that fail to respond to antisecretory therapy also require additional evaluation.

Indications for Surgery and Management

Indications for surgical management of GERD include suboptimal symptom control with nonoperative treatment, complicated GERD with stricture or bleeding, and patient preference for cessation of pharmacologic agents. A thorough preoperative anatomic and physiologic evaluation is important to both clearly establish the diagnosis and to aid in selecting the most appropriate operative procedure.

Considerations that affect the choice of surgical approach (laparoscopic or open) and the specific operation performed include the presence of a large hiatal hernia, short esophagus, substantially impaired esophageal peristalsis, morbid obesity, and history of prior upper gastrointestinal operations. Endoscopic antireflux options are emerging for select patients with GERD. However, the techniques and instrumentation are not yet fully developed and long-term efficacy has not been established.

The principle of an antireflux operation is to obtain a higher pressure segment of intra-abdominal esophagus. The standard operation for most patients is a laparoscopic Nissen fundoplication, which creates a 2–3 cm long 360° (complete) wrap of gastric fundus around the distal esophagus. Adequate mobilization of the fundus is important to prevent the wrap from being too tight, which will result in postoperative dysphagia. The vagus nerves are preserved. The diaphragmatic crura are approximated if the hiatus is patulous. The Nissen fundoplication has an 85–90 % success rate for control of GERD symptoms.

Surgical options for antireflux procedures include transabdominal fundoplication. The laparoscopic Nissen fundoplication, often with the Rossetti modification, is the most commonly

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_33, © Springer New York 2015

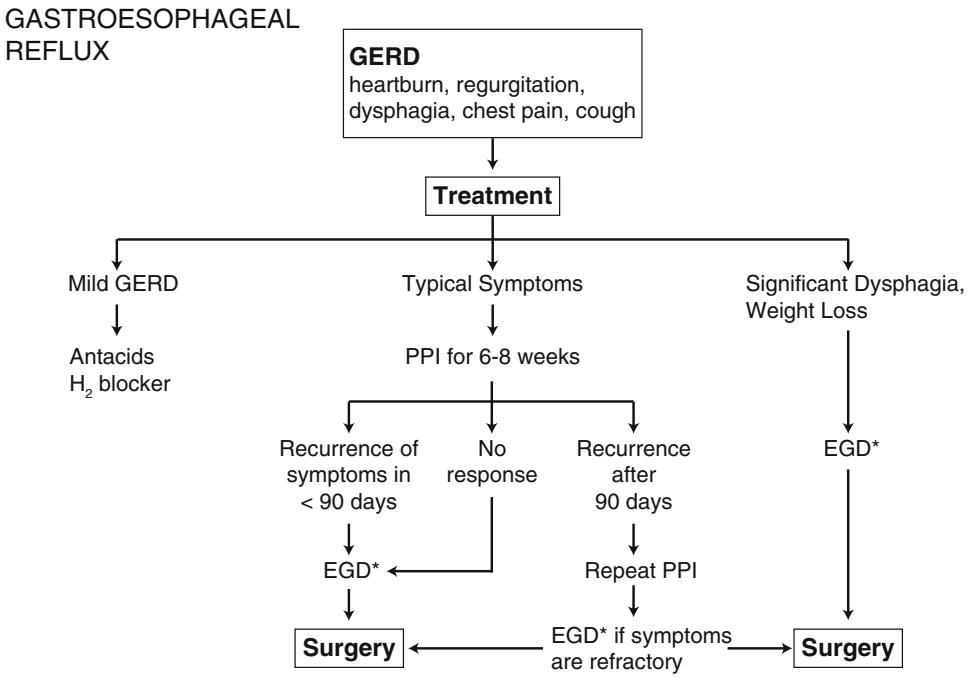

FIG. 33.1 Treatment algorithm for gastroesophageal reflux disease (GERD). *PPI* proton pump inhibitor, *EGD* esophagogastroduodenoscopy, *UGI* upper gastrointestinal

used and has shown long-term success rates on the order of 80–90 %. Complications of fundoplication may include dysphagia (10 %), gas bloat syndrome (20 %), and up to 30 % of patients may require the addition of an antihistamine or proton pump inhibitor (PPI) after long-term follow-up.

For patients with a component of esophageal dysmotility, the surgeon may consider a partial wrap, such as the Dor or Toupet. A Belsey Mark IV is also a reasonable transthoracic approach with acceptable results. Surgical options for patients with atypical findings may include a Collis gastroplasty or a Hill esophagogastropexy. Findings of high-grade dysplasia on esophagogastroduodenoscopy (EGD) will require either endoscopic mucosal resection or esophagectomy. Malignancy is dealt with accordingly.

There are various types of partial fundoplications that can be used in selected circumstances. For example, when concomitant esophagomyotomy or paraesophageal hernia repair is done or when esophageal peristalsis is deficient. These partial fundoplications include the Toupet posterior fundoplication, Dor anterior fundoplication, Hill gastropexy, and Belsey Mark IV transthoracic fundoplication.

34
Acute Pancreatitis

Hadyn Hollister and Keith W. Millikan

Presentation

The cardinal symptom of acute pancreatitis is *severe abdominal pain*. It is typically epigastric in location with radiation through to the mid-central back, although it may involve any part of the abdomen or lower chest; it is sharp in quality, rapid in onset, and unremitting. Patients will often shift positions in search of greater comfort. Nausea, intractable vomiting or retching, and anorexia often accompany. On abdominal examination, patients will exhibit varying degrees of tenderness, which may be localized to the epigastrium or may be diffuse, depending upon the severity of the attack. In developed countries, 70–80 % of acute attacks are associated with *alcohol abuse* or *choledocholithiasis* (Fig. 34.1). Accounting for 10–15 % of cases are hypertriglyceridemia, hypercalcemia, medications, external ductal obstruction, endoscopic retrograde cholangiopancreatography (ERCP), operative insult, trauma, infection, ischemia, and scorpion venom. Hereditary conditions, metabolic disorders, and autoimmune diseases can also lead to bouts of acute pancreatitis. The remaining 10–15 % of cases are idiopathic in etiology. In patients with a high clinical suspicion for acute pancreatitis, *the diagnosis is typically established via laboratory findings of hyperamylasemia greater than three times the upper limit of normal or hyperlipasemia, which is the more specific measurement of the two*. Ultrasound may be utilized to identify gallbladder stones, dilatation of the biliary and pancreatic ductal systems, or a diffusely enlarged and edematous pancreas, although findings may be obscured by the presence of intestinal gas. While computed tomography with intravenous contrast should not be ordered reflexively as part of the workup for acute pancreatitis, it may be of use when the diagnosis of acute pancreatitis is not evident from clinical or laboratory findings and should not be withheld when the clinical picture is suggestive of a severe attack.

Evaluation of Severity

Early discernment of the severity of an attack of acute pancreatitis is of utmost importance in deciding upon an appropriate pathway of treatment. Cases may be divided into two broad categories: edematous or mild acute pancreatitis, which accounts for approximately 90 % of attacks, and necrotizing or severe acute pancreatitis. Morbidity and mortality are significant in the latter and far less common in the former. Commonly used scoring systems include *Ranson's criteria* (≥3 points signifying a severe attack) (Table 34.1) and the Acute Physiology and Chronic Health Evaluation II (*APACHE-II*) scoring system (≥8 signifying a severe attack). A CT severity index correlating morbidity and mortality with the radiographic appearance of the pancreas and the presence and degree of necrosis has been developed by Balthazar et al. (Table 34.2). Patients in shock or evidencing organ failure are managed as severe cases; tachycardia, tachypnea, and hypotension will be evident in such patients. A serum C-reactive protein (CRP) >150 mg/dl at 48 h is indicative of a severe attack.

Initial Management: Mild Acute Pancreatitis

Patients with mild acute pancreatitis should be managed conservatively with fluid resuscitation, electrolyte replacement, nothing per mouth until there is symptomatic improvement, and IV pain medication. If the patient is vomiting, a nasogastric tube should be placed. There has been no observed benefit for patients in whom prophylactic antibiotics are used in mild attacks. A CT study should not be considered a routine element of the workup in patients who are not severely ill. As a general rule, it should only be performed in such patients if

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_34, © Springer New York 2015

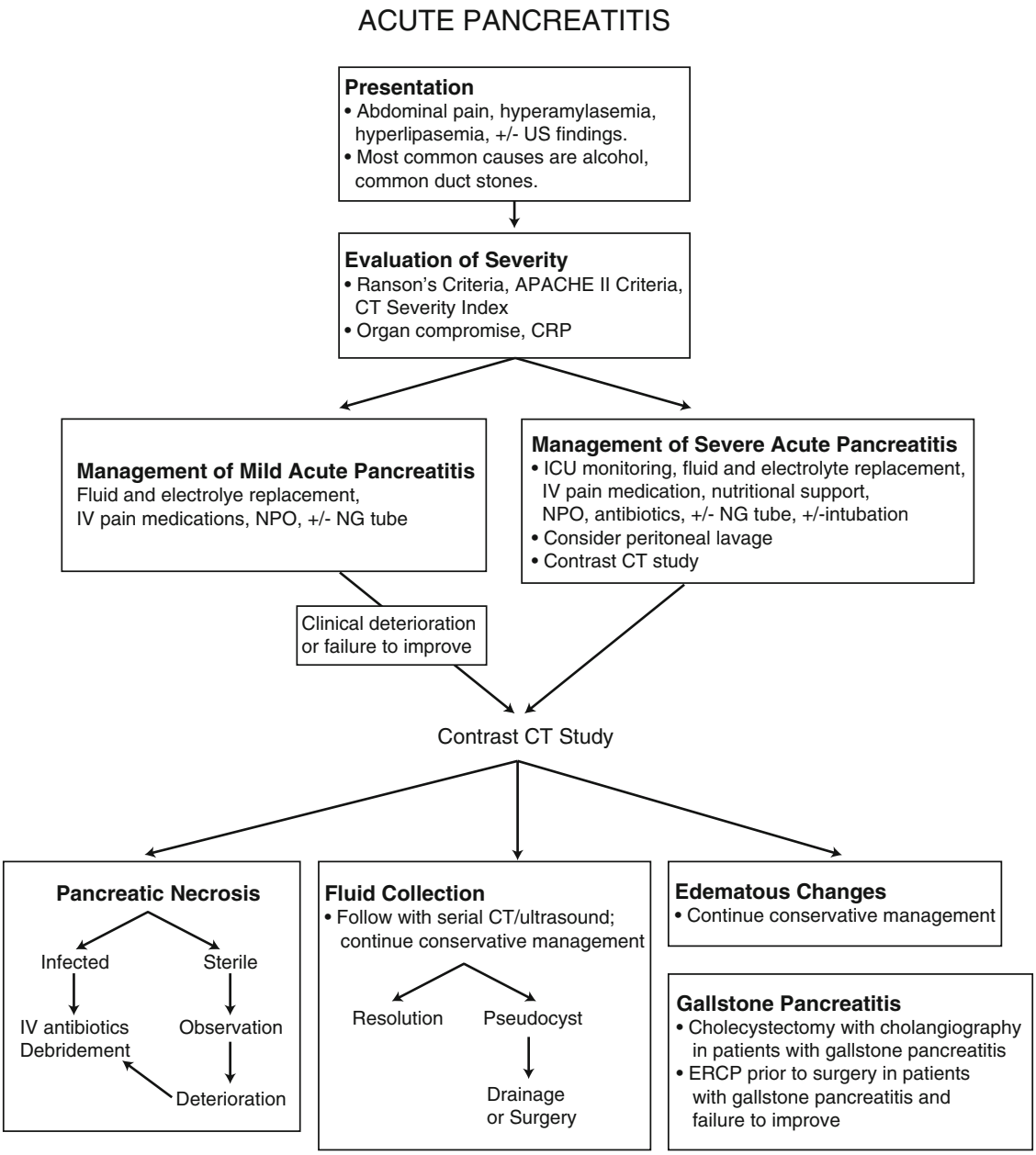

FIG. 34.1 Treatment algorithm for pancreatitis. *US* ultrasound, *APACHE II* Acute Physiology and Chronic Health Evaluation II, *CT* computed tomography, *CRP* C-reactive protein, *IV* intravenous, *NPO* nothing per oral, *NG* nasogastric, *ICU* intensive care unit, *ERCP* endoscopic retrograde cholangiopancreatography

TABLE 34.1. Ranson's criteria.

Criteria present upon admission	Criteria present within 48 h of admission	Mortality by total criteria present
Age over 55 years	Hematocrit drop greater than 10 %	0–2: 0–3 %
White blood cell count over 16,000/μ(mu)L	Blood urea nitrogen rise greater than 5 mg/dL	3–5: 11–15 %
Blood glucose over 200 mg/dL	Arterial PO_2 of less than 60 mmHg	6–11: ≥40 %
Serum lactate dehydrogenase over 350 units/L	Serum calcium less than 8 mg/dL	
Aspartate aminotransferase over 250 units/L	Base deficit greater than 4 mEq/L	
	Fluid sequestration greater than 6 L	

TABLE 34.2. CT severity index.

Prognostic indicator	Points	Morbidity/ mortality by total points
Pancreatic inflammation		
A: Normal pancreas	0	0–3: 8 %, 3 %
B: Focal or diffuse enlargement	1	4–6: 35 %, 6 %
C: Intrinsic pancreatic abnormalities with inflammatory changes in peripancreatic fat	2	7–10: 92 %, 17 %
D: Single, ill-defined fluid collection or phlegmon	3	
E: Two or more poorly defined collections or presence of pancreatic or peripancreatic gas	4	
Pancreatic necrosis		
None	0	
<33 %	2	
33–50 %	4	
>50 %	6	

there is an absence of clinical improvement within the expected time frame of 5–7 days, if there is overt deterioration, or if there is reason to believe that a ductal obstructing tumor may be responsible for the attack.

Initial Management: Severe Acute Pancreatitis

Patients with severe acute pancreatitis should be admitted to an intensive care unit, receive aggressive fluid resuscitation and electrolyte replacement, receive nothing by mouth, and be treated with IV pain medication. A nasogastric tube should be placed if the patient is vomiting, and intubation may be necessary if there is respiratory compromise. Heart rate, blood pressure, oxygen saturation, and urine output should be closely monitored; measurement of central filling pressures should be considered. There is some debate as to whether it is appropriate to administer IV antibiotics to patients suffering severe attacks. Studies have shown some benefit with regimens including imipenem alone, imipenem with cilastatin, and cefuroxime. Some argue that antibiotics should never be given prophylactically because of the inherent risks of selection for resistant organisms and fungal superinfection. Others advocate the cautious administration of antibiotics along with an antifungal agent. It is this author's opinion that broad-spectrum antibiotics should be administered in cases of severe acute pancreatitis, as this will generally delay the need for surgery, thereby allowing the patient to be clinically optimized and improving postoperative outcomes. The role of *peritoneal lavage* in severe attacks is also the subject of some debate, with early studies demonstrating improvement in early mortality and more recent studies demonstrating diminished levels of inflammatory factors both within the pancreas and the serum. However, meta-analyses have thus far failed to demonstrate a definitive advantage, and as such, the use of lavage should be dependent upon surgeon preference and individual patient situation. Because patients with severe acute pancreatitis are typically unable to eat for prolonged periods of time, *nutritional support should be provided* within at least 72 h of hospitalization. Total parenteral nutrition through a central line can be utilized if patients are not able to tolerate small amounts of enterally administered feeds via nasogastric (NG) or nasojejunal (NJ) tube. This latter approach confers the theoretical benefit of preventing bacterial translocation via trophic effects exerted upon the injured bowel wall abutting the inflamed pancreas, thereby decreasing the incidence of pancreatic infection. Finally, as noted previously, CT imaging should be obtained in all patients suffering a severe attack to evaluate for necrosis, infection, and fluid collection.

Pancreatic Necrosis

Patients with evidence of necrosis on CT are managed differently depending upon whether or not there is a high degree of suspicion for infected necrosis. This may be suggested by the appearance of air pockets on CT as well by the patient's clinical status. When infected necrosis is suspected, antibiotic therapy should be initiated. Percutaneous CT-guided aspiration should be considered; whether this is performed immediately or only after the patient has failed to improve clinically after a trial of antibiotics is a matter of physician preference and specific patient scenario. Finally, it is generally agreed upon that surgical debridement is appropriate in cases of infected necrosis. Laparotomy with debridement is the conventional procedure, although recent studies have found improved outcomes in patients managed with percutaneous drainage, an endoscopic transpapillary approach, or laparoscopic retroperitoneal debridement, while reserving laparotomy for patients in whom more conservative interventional measures fail. Patients with evidence of necrosis on CT in whom suspicion for infected necrosis is not high (or in whom aspiration fails to identify bacterial organisms) should be managed conservatively. If there is clinical deterioration or failure to improve, antibiotic therapy should be initiated and aspiration should be performed.

Fluid Collection

Thirty to fifty percent of patients will develop acute fluid collections during the early stages of an attack. These simple collections are not walled off by fibrinous or granulation tissue and a majority will resolve spontaneously. Those that remain may develop into pancreatic pseudocysts. Because acute fluid collections will regress without intervention in a majority of cases, they should be followed with ultrasound or CT scan until resolution. If the collection becomes walled off by fibrinous or granulation tissue, grows to become larger than 5 cm in diameter, or is persistent for a prolonged period of time (generally, periods in excess of 4–6 weeks), drainage (percutaneous or endoscopic) or surgical excision should be performed.

Edematous Changes

Patients with edematous changes on CT should be managed conservatively as described above in "Mild Acute Pancreatitis".

Additional Considerations

In addition to following the management steps outlined previously, the underlying pathology should in every case be elucidated and appropriately rectified. Patients found to have gallstone pancreatitis on ultrasound or CT should undergo cholecystectomy with cholangiography during the same hospitalization. Preoperative ERCP with sphincterotomy should be considered, particularly for patients with gallstone pancreatitis in whom clinical and laboratory improvement is not evident. However, ERCP should not be performed as a matter of routine, as a minority of patients will have choledocholithiasis at time of surgery. Management options for patients found to have retained stones during surgery include immediate common bile duct exploration with stone extraction or postoperative ERCP. Patients with gallstone pancreatitis in whom clinical and laboratory improvement are not evident should undergo ERCP with sphincterotomy and stone extraction prior to cholecystectomy. Finally, when investigation fails to uncover an etiology for acute pancreatitis, ERCP should be performed to rule out the presence of a ductal obstructing neoplasm.

35
Chronic Pancreatitis

Joseph Broucek and Jonathan A. Myers

Etiology and Pathogenesis

Long-term alcohol abuse is the most common cause of chronic pancreatitis in westernized nations, with a reported incidence of 3.5–10 per 100,000. Other causes include high-fat and protein diets, untreated hyperparathyroidism with hypercalcemia, ductal obstruction (i.e., strictures, cancer, stones), autoimmunity, genetic mutations (i.e., CFTR), tropical, or idiopathic. The primarily accepted pathogenesis is that of necroinflammation and fibrosis that is recurrent and progressive in nature (Fig. 35.1). Inciting or recurring events trigger and potentiate the disease, as seen in alcohol abuse-related pancreatitis. This may lead to organ dysfunction, pain, and eventual burnout. This disease is associated with a 50 % mortality rate within 20–25 years, although only 15–20 % is known to die of an attack of pancreatitis. Death is usually due to other comorbidities or factors associated with alcohol abuse (i.e., tobacco abuse, trauma, malnutrition, or infections).

Presentation and Physical Examination

Chronic pancreatitis usually presents with *continuous, daily pain*, which may require analgesics or prompt increased alcohol ingestion for relief. Occasionally, exocrine and/or endocrine insufficiency are the first signs of chronic pancreatitis, but these usually become manifest after pancreatitis has been present for a long duration. A pseudocyst, obstruction of the pancreatic duct, or replacement of the gland by a diffuse fibrocalcific process may cause pain by involving the celiac plexus. Steatorrhea may also be present due to loss of pancreatic function; however, 80–90 % of exocrine and endocrine function must be lost before these symptoms are seen. Since smoking is a habit that frequently accompanies alcohol abuse, patients may have chronic obstructive pulmonary disease. If impairment of the gland exists, then one may often have what is classified as type III diabetes secondary to loss of both insulin- and glucagon-producing cells. Ductal obstruction at the pancreatic head (due to stricture, stones, etc.) can lead to jaundice as well.

Physical examination is, for the most part, unremarkable. If pain is present, it is most typically located in the epigastric region, is sharp in nature, and is described as radiating to the back or scapula. This pain can be relieved by leaning forward and is worsened with eating. Ten to twenty percent will not have pain. There may also be signs of malnutrition as food stimulates pancreatic enzyme release, thereby worsening pain. Consequently, patients are afraid to eat and significant weight loss is apparent.

Diagnosis

Other causes of upper abdominal pain must be excluded; tests that assist in this process include upper gastrointestinal endoscopy to rule out peptic ulcer disease and an ultrasound to rule out cholelithiasis. A plain radiograph of the abdomen will reveal pancreatic calcifications in 30–40 % of patients and can be useful in the setting of steatorrhea to aid in making the diagnosis of chronic pancreatitis. Pancreatic function tests may help delineate an acute attack as well as assess the overall exocrine and endocrine function of the gland. *The presence of normal serum amylase and/or lipase does not exclude the diagnosis of chronic pancreatitis*. When ductal obstruction is present, elevations of alkaline phosphatase and serum bilirubin can be seen.

The pancreas is usually not well visualized by transabdominal ultrasound, although one may visualize calcifications, ductal dilation, and/or cysts. A computed tomography (CT) scan of the abdomen is generally recommended and may demonstrate edema, acute inflammation, necrosis (during a recurrent attack), as well as signs of chronic changes such as fibrosis, inflammation, tissue loss, ductal dilation, calcifications, and a pseudocyst. To image the duct, endoscopic retrograde cholangiopancreatography (ERCP)

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_35, © Springer New York 2015

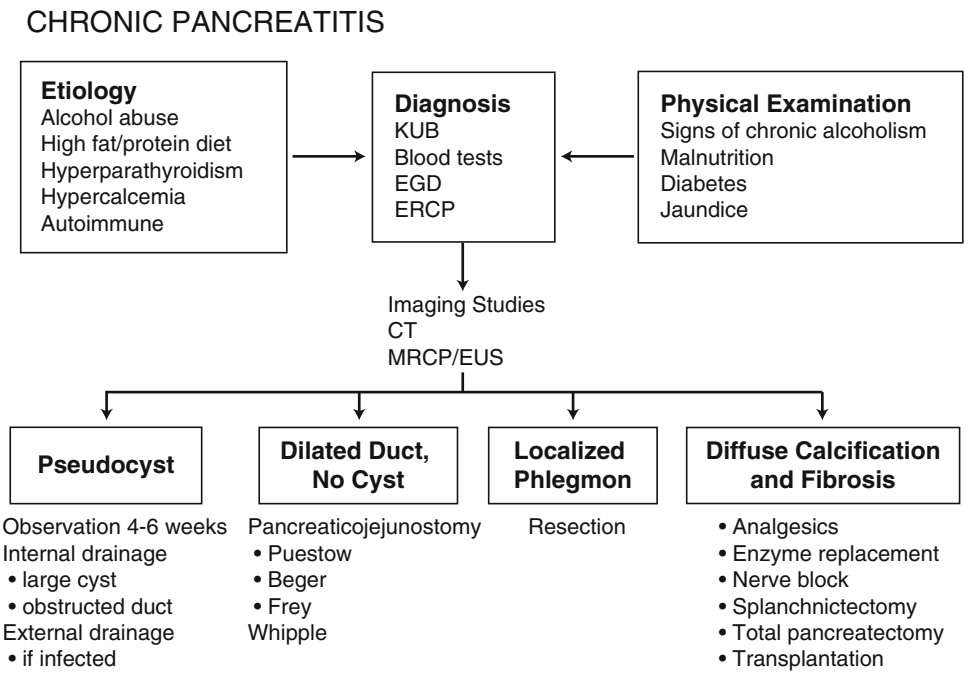

FIG. 35.1 Chronic pancreatitis treatment algorithm. *KUB* kidneys, ureters, bladder, *EGD* esophagogastroduodenoscopy, *ERCP* endoscopic retrograde cholangiopancreatography, *CT* computed tomography, *MRCP* magnetic resonance cholangiopancreatography, *EUS* endoscopic ultrasound

can be performed; this may be useful if biliary obstruction (30 % of pancreatitis) is also suspected. ERCP could also identify dilation, stenosis, cysts, and abnormalities of the side branches. While ERCP is considered the gold standard in imaging, it can be technically difficult and the endoscopist may not be able to cannulate the pancreatic duct. In this situation, MRCP (magnetic resonance cholangiopancreatography) or endoscopic ultrasound can be performed. These modalities are very sensitive in detecting the disease in its later stages.

Duodenal obstruction is present in 10–15 % of cases and may preclude successful ERCP. In these instances, an upper gastrointestinal (UGI) barium exam may reveal a stricture usually in the second portion of the duodenum. Tumor markers (Ca 19-9, CEA) should be obtained if an occult malignancy is suspected.

Pseudocysts

These are defined as collections of secretions from the pancreas that are lined by non-epithelial fibrous walls composed of granulation tissue. They develop in about 10 % of patients with chronic pancreatitis, and two-thirds originates in the body or tail. If pain is present in chronic pancreatitis, one should suspect a pseudocyst. If a pseudocyst is found in conjugation with chronic pancreatitis, observation for a period of 4–6 weeks may allow the cyst wall to either mature or spontaneously resolve. For simple pseudocysts without proximal ductal obstruction, an ERCP endoscopist can first attempt endoscopic internal drainage. The success rate associated with this approach is approximately 80 % with a recurrence rate of 10–20 % and a mortality rate of up to 3 %. If unsuccessful, surgery should be considered. Cysts that are symptomatic, persist, enlarge, are greater than 5 cm, and/or are associated with proximal ductal obstruction may be drained internally into the stomach, duodenum (if the cyst is adherent), or a limb of jejunum via a Roux-en-Y reconstruction, depending on the location of the cyst. External drainage is preferred only when a mature cyst wall is lacking or if the cyst has become an abscess. In the appropriate patient, distal pancreatectomy may be considered for cysts in the tail of the pancreas; however, caution should be exercised since extensive loss of tissue could impair pancreatic exocrine and endocrine function.

Dilated Duct, No Pseudocyst

For the patient with severe, unrelenting pain and a pancreatic duct that is dilated greater than 7 mm, there are three common drainage procedures. The most popular is the *modified Puestow* procedure, which is a lateral pancreaticojejunostomy; this drains the main pancreatic duct into a Roux limb of jejunum over a distance of at least 10 cm. This procedure relieves pain in 80 % of patients. The *Beger* procedure is popular in Europe

and consists of a duodenal-preserving resection of the head of the pancreas and drainage of the body and tail duct and a small rim of pancreatic head along the duodenum into a Roux limb. The success rates obtained in Europe with the Beger procedure have not been duplicated in the United States. The *Frey* procedure is a modification of the Puestow and consists of coring out the head of the pancreas in conjunction with a lateral pancreaticojejunostomy extended onto the tail. This procedure combines both the Puestow and the Beger and is gaining popularity in the United States. If biliary (30 %) and/or duodenal (15 %) obstruction is present, choledochoduodenostomy or choledochojejunostomy and/or gastrojejunostomy may be required. Some surgeons will perform a pyloric-preserving pancreaticoduodenectomy with a Puestow procedure of the body and tail when biliary and/or duodenal obstruction exists.

Localized Phlegmon

If the workup reveals a localized inflammatory process of either the head or the body or the tail of the pancreas, a resection of the localized area is the appropriate therapy. For an inflammatory mass of the head of the pancreas, a pyloric-preserving pancreaticoduodenectomy is recommended. For an inflammatory mass of the body of tail of the pancreas, a distal pancreatectomy with splenectomy is performed.

Normal Duct, Fibrotic Gland

The most common symptom necessitating treatment is pain. *Abstinence from alcohol or other inciting agent is the first step in treatment*. For a diffusely fibrocalcific pancreas with a normal-sized duct, the next line of therapy includes *analgesics and pancreatic enzyme replacement* to enhance negative feedback on the pancreas and oppose cholecystokinin-mediated stimulation. If this is not successful, *a celiac nerve block* can be performed through a percutaneous injection of phenol or alcohol. These blocks relieve pain for only a period of 6 months and have been used mainly for patients with pancreatic cancer. *Splanchnicectomy* has become popular in France with very promising results that have not been duplicated in the United States. The procedure most commonly performed for a diffusely fibrocalcific pancreas with a normal-sized duct is *near total pancreatectomy*. Its disadvantage is that it causes exocrine and endocrine insufficiency, namely, brittle diabetes and its complications. Some authors believe that near-total pancreatectomy is overly radical and that the pancreas should be allowed to burn itself out (the pain eventually resolves). *Pancreatic autotransplantation* has been attempted but has many complications, including moving the pain and the location of pancreatitis to the site of autotransplantation. Islet cell transplants have also been attempted, but two to three pancreas organs are required for successful islet function. A limited supply of transplantable pancreas organs has kept this procedure from gaining wide acceptance. Although *endoscopic* stenting of the pancreatic duct has been shown to relieve symptoms in up to 30–76 % of patients, it is considered a short-term solution and not a definitive therapy.

Pancreatic Leak, Pleural Fistulas, and Other Complications

Should a pseudocyst or duct rupture, pancreatic fluid may leak causing free fluid in the abdomen or even create a fistula to the pleural space. This is diagnosed by sampling the fluid (abdominal and thoracic) and documenting high levels of amylase and protein. A conservative approach to treatment may be considered including serial paracentesis (resolution in 50–60 % of patients). One may also use diuretics, octreotide, and carbonic anhydrase inhibitors in an effort to decrease fluid leak. Endoscopic intervention can be performed including pancreatic sphincterotomy and/or stent placement in hopes of bridging the leak and allowing anterograde drainage into the duodenum. More commonly, surgery may be performed including mobilizing a loop of jejunum for a Roux-en-Y anastomosis to the site of rupture if more proximal. If the site of rupture is distal, a distal pancreatectomy is an option. Other complications of inflammation and pseudocysts may include the formation of false aneurysms, pancreaticoenteric fistulas, and splenic vein thrombosis leading to varices.

36
Pancreatic Cancer

John Christopher McAuliffe and John D. Christein

Epidemiology

Pancreatic cancer (PC) is the *fourth leading cause of cancer-related mortality* in the United States and annually up to 230,000 people worldwide will die of PC. Approximately 43,000 will be diagnosed with PC in the United States this year, making it second only to colorectal cancer in tumors of gastrointestinal origin. The estimated overall 1-year survival rate is only 22 % with less than 5 % survival at 5 years—figures that underscore PC's aggressive nature and the need for early detection. *Surgical resection is the only potentially curative modality*; however, even with advances in cross-sectional imaging and endoscopic diagnostics, only 15–20 % of patients will have resectable disease at presentation.

Risk factors include smoking, history of chronic pancreatitis, advanced age, male sex, diabetes mellitus, obesity, non-O blood group, chlorinated hydrocarbon solvents and nickel exposure, African American race, high-fat diet, and perhaps *Helicobacter pylori* infection. The majority of PC is sporadic, while well *less than 10 % of patients have a family history*. Specific genetic aberrations linked to PC include KRAS, CDKN2A, TP53, SMAD4, STK11, PRSS1, PALB2, BRCA2, and telomere shortening. Similar to other gastrointestinal carcinomas, PC proceeds through a consistent histologic pathway from normal to hyperplasic epithelium (self-sufficiency in growth signals and insensitivity to anti-growth signals), evasion of apoptosis, autonomous replication, angiogenesis, tissue invasion, and, finally, metastasis. Molecular evolution studies of PC show that a neoplastic clone will take more than 10 years to become a malignancy, providing potential opportunities for screening and early diagnosis, of which none have been developed.

History and Physical Exam

Early-stage disease is typically silent and rarely found. Early tumors may be found incidentally during evaluation of unrelated symptoms (Fig. 36.1). The disease becomes apparent after causing pancreatic endocrine or exocrine dysfunction, invasion into adjacent structures causing obstruction of the biliary or upper gastrointestinal tract, or symptoms from metastatic disease. Most patients who present with symptoms have locally advanced or metastatic disease. Approximately 80 % of PCs are located in the pancreatic head and cause painless jaundice, mid-back or epigastric pain, and weight loss. Other symptoms may include anorexia, new-onset diabetes, steatorrhea, alcoholic stools, pancreatitis, gastric-outlet obstruction, and/or deep venous thrombosis. Up to 60 % of patients present with glucose intolerance or diabetes mellitus many months prior to a tumor or mass symptoms. No physical finding is pathognomonic. Patients may appear cachectic, jaundiced, have lymphadenopathy, mass effect in the epigastrium or right upper quadrant, and lower extremity swelling secondary to deep venous thrombosis.

Diagnostic Evaluation

Triphasic (arterial, late arterial, and venous) pancreatic-protocol computed tomography (CT) is the single best initial diagnostic test for pancreatic cancer. Triphasic CT also accurately evaluates stage and resectability. Contrast enhancement between normal, adjacent tissue and the hypodense, fibrotic PC is highest during the late arterial phase. Chest radiography is required to determine if pulmonary metastases are present. Positron emission/CT is not part of the routine workup for PC but can be used to evaluate for the presence of CT-negative disease, metastatic disease, or recurrence after resection. Magnetic resonance with gadolinium enhancement is used for those intolerant to iodine contrast used in CT.

Histologic diagnosis is not required before surgery when a patient has resectable disease and is a fit operative candidate. However, a histologic diagnosis will often be required in the presence of locally advanced or borderline resectable PC or metastatic disease before beginning chemotherapy.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_36, © Springer New York 2015

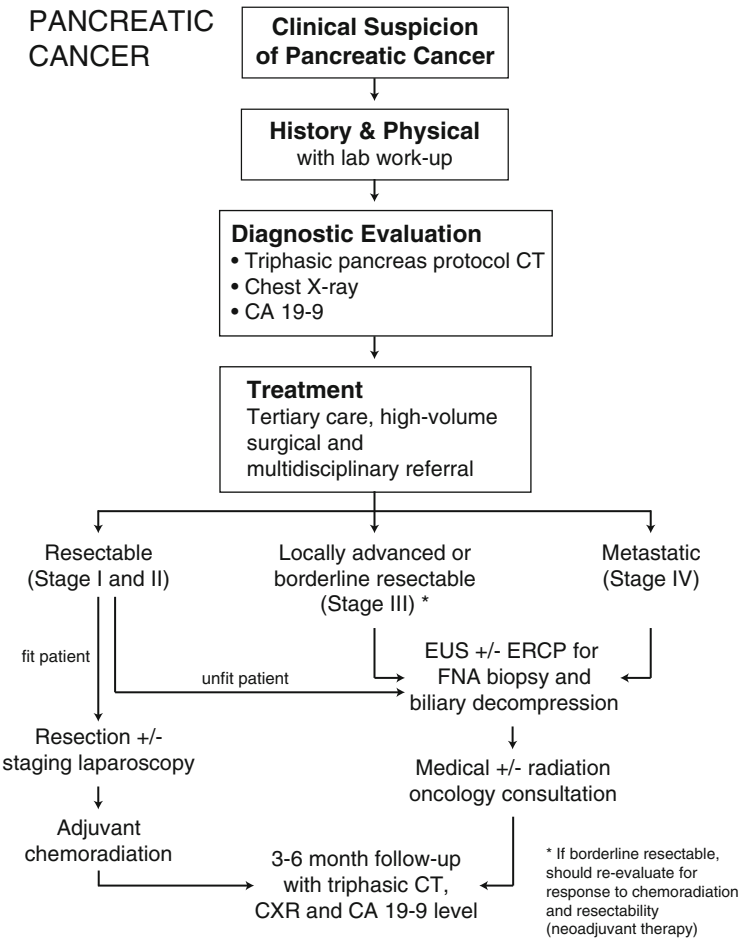

FIG. 36.1 Treatment algorithm for pancreatic cancer. *CT* computed tomography, *EUS* endoscopic ultrasound, *ERCP* endoscopic retrograde cholangiopancreatography, *FNA* fine-needle aspiration, *CXR* chest X-ray

Endoscopic ultrasound (EUS) and/or endoscopic retrograde cholangiopancreatography (ERCP) can be used for tissue diagnosis and biliary decompression, respectively.

Treatment

Patients with proven PC or a suspicious pancreatic mass should undergo evaluation at a tertiary care, high-volume center with experienced pancreatic or hepatobiliary surgeons. High-volume centers have improved perioperative outcomes and the necessary support staff and multiple specialties to treat complications from pancreatic operations. Based on cross-sectional imaging, patients are initially stratified to resectable (stage I and II), borderline or locally advanced unresectable (stage III), or metastatic disease (stage IV).

Fit patients with resectable (stage I or II) disease should undergo resection. Some surgeons prefer preoperative biliary decompression prior to resection; however, this is controversial. For those with stage III or IV disease, neoadjuvant or palliative therapy is considered first, resection is not frontline therapy for these patients. For patients of poor operative status or those about to receive neoadjuvant therapy, biliary decompression with ERCP and stent (metallic or plastic) should be considered. Patients who present with metastatic disease should be considered for metallic stent placement.

For those with radiographically resectable disease and high-risk features such as size >2 cm, location in the body or tail, vascular abutment, or suspicious distant lesions, some advocate *staging laparoscopy* with intraoperative frozen section. This may identify patients who are in fact unresectable, thereby sparing them an unnecessary laparotomy.

Pancreaticoduodenectomy, the Whipple procedure, is the operation of choice to treat cancer of the head of the pancreas as well as other periampullary malignant and premalignant lesions. For tumors of the pancreatic body or tail, distal pancreatectomy and splenectomy should be performed (perhaps proceeded by staging laparoscopy). To date, no randomized trial has proven that the laparoscopic approach is equivalent to open operation for pancreatic cancer. The median survival after complete resection can be up to 20 months with 5-year survival rates approaching 25 %. The 90-day complication rate is 60 % following the Whipple operation; 20 % of complications are serious and require additional intervention. Marked gains in mortality have been achieved with rates

lower than 5 % noted at high-volume centers. The most common and characteristic complication (20 %) is a pancreatic fistula. Other complications include pneumonia, wound complications, delayed gastric emptying, and anemia.

Postoperatively, all patients should be evaluated for adjuvant therapy. Once patients have recovered from surgery, they should undergo triphasic, pancreas protocol CT imaging to evaluate for residual or metastatic disease. Additionally, CA 19-9 levels should be drawn at this time. Adjuvant chemotherapy has shown efficacy, but no standard protocol exists for chemotherapy and/or radiation. During the past 10 years, gemcitabine-based regiments appear to have the most efficacy. Immunologic therapies are also being studied. Those with poor performance status are not candidates for systemic therapy and should be provided optimal supportive care.

Frontline systemic postoperative adjuvant therapy includes gemcitabine followed by 5-fluorouracil-based chemoradiation or chemotherapy alone (gemcitabine or 5-flourouracil/leucovorin or capecitabine). Frontline systemic therapy for those with stage III or IV disease is similar. With regard to those with stage III disease, systemic treatment may produce enough tumor shrinkage to render lesions resectable. Therefore, imaging studies should be obtained periodically to assess tumor response.

Patients undergo surveillance every 3–6 months for 2 years, then annually. At each follow-up, a CA 19-9 serum level and triphasic CT is obtained.

Conclusion

Considerable progress has been made in the evaluation and treatment of patients with PC. Despite this, survival is poor and therapy is morbid. A better understanding of the pathophysiology of PC will provide the rationale for more efficacious therapies required for this disease. Using current practice guidelines, high-volume institutions with multidisciplinary management provide the best chance at cure and care of patients and families with PC.

37
Small Bowel Obstruction

David D. Shersher

Introduction/Etiology

Despite advancement in medical and surgical techniques, small bowel obstruction (SBO) continues to be a challenge for general surgeons. Although hernias were the most common etiology of SBO a century ago, postoperative adhesions now account for at least 75 % of small bowel obstructions (Fig. 37.1). Additional etiologies of obstruction include tumors and less commonly, Crohn's disease, volvulus, intussusception, gallstone ileus, and bezoar.

Laparoscopy was initially predicted to diminish small bowel obstruction from adhesions, but this is debatable. Nearly 25 % of obstructed patients require an operation sometime during their index presentation.

Operative versus nonoperative management strategies have been compared with respect to recurrence of SBO; one in three surgically treated patients recur compared to one in two medically managed patients. Despite this, many patients are successfully managed nonoperatively, albeit with repeat emergency department visits.

Initial Evaluation

The initial evaluation of a patient with suspected SBO is a thorough history and physical examination. Symptoms often include nausea and vomiting, abdominal distention, crampy abdominal pain, and obstipation. A proximal small bowel obstruction often presents with bilious emesis compared to a distal obstruction that may present with feculent vomiting. Previous abdominal surgeries are important contributors to adhesive bands; the absence of a prior surgical history may be an indicator to operate sooner. Additionally, it is important to inquire about personal and family history of colorectal cancer and inflammatory bowel disease as well as the date of the last colonoscopy.

Physical examination begins with vital signs; signs of tachycardia and/or hypotension may alert the examiner to dehydration or sepsis. Abdominal exam usually reveals distention, tympany, and high-pitched or absent bowel sounds on auscultation. Examination for hernia is critical. Patients with evidence of bowel ischemia or perforation often present with tenderness, rigidity, and guarding indicative of peritonitis.

The laboratory exam is often nonspecific, but patients who present with vomiting may have a hypochloremic, hypokalemic metabolic alkalosis with paradoxic aciduria. Additionally, leukocytosis may indicate an inflammatory or infectious process. Acidosis is usually associated with bowel ischemia or perforation causing peritonitis.

Initial plain X-ray films should be done in the supine, upright, and lateral decubitus views to evaluate for pneumoperitoneum, bowel dilation (>3 cm), air-fluid levels, and a paucity of colon or rectal gas. If plain films are nondiagnostic, computed tomography (CT) is used to elucidate the location and degree of obstruction. CT can also discover a mass or hernia and is particularly helpful in finding intra-abdominal abscess if done with intravenous and oral contrast. In patients with persistent SBO, a small bowel follow-through—CT or magnetic resonance enterography—may help define a point of luminal stricture; however, this should be avoided if one suspects a complete obstruction.

Nonoperative Management

Nearly 75 % of partial obstructions resolve with nonoperative management consisting of intravenous fluids, nasogastric decompression, and nothing per os (NPO). Intake and output should be monitored; a urinary indwelling catheter may facilitate this. Serial abdominal examinations are performed along with daily X-ray films. Unless a patient had a recent abdominal surgery, failure to symptomatically improve with nonoperative management after 24 h requires consideration for surgical exploration. Conversely, patients who demonstrate an improving exam may be challenged with a judiciously advanced diet and be sent home after passage of flatus and stool.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_37, © Springer New York 2015

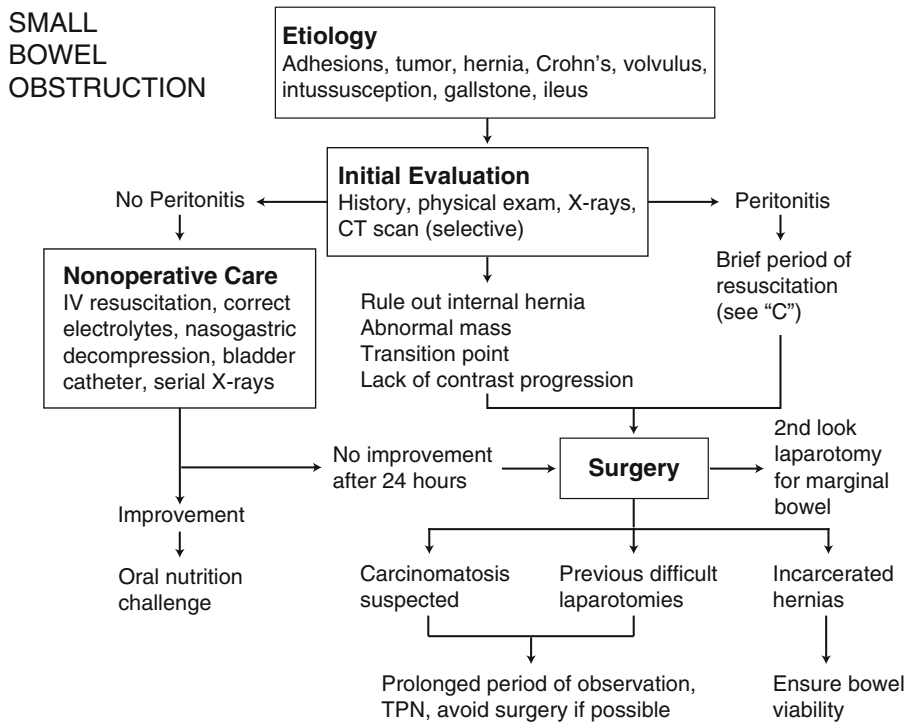

FIG. 37.1 Algorithm for treatment of small bowel obstructions. *CT* computed tomography, *IV* intravenous, *TPN* total parenteral nutrition

Operative Management

Twenty-five percent of patients with SBO will require an operation. Prompt operative intervention is required for patients with peritonitis or evidence of perforated viscus or bowel ischemia. If a patient with an SBO has never had a previous abdominal operation nor a hernia on exam, consideration for surgical exploration should be prioritized. Surgery should also be strongly considered for patients who have no clinical improvement after 24 h of nonoperative management; many of these patients will simply require adhesiolysis, but some may need resection for nonviable bowel or a tumor.

A history of intra-abdominal malignancy poses a special challenge as two-thirds of these patients have SBO from adhesions from previous surgery and one-third from carcinomatosis. Unless there is clear evidence of carcinomatosis on imaging studies, these patients should undergo surgery. Many would argue that even in the presence of carcinomatosis, surgery should be offered. Relief of the obstruction might be possible and is preferable to life without the pleasure of being able to eat. Intussusception in adults is also challenging; one-half is caused by a neoplastic lead point and may need eventual surgical resection if they do not resolve with conservative management.

Adhesive small bowel obstructions from numerous abdominal operations are frequently technically difficult to manage due to loss of tissue planes and distorted anatomy. Unless a patient has clearly failed medical treatment, surgery should be deferred until after a long nonoperative course of NPO and total parenteral nutrition (TPN).

Hernias that are incarcerated, but not strangulated, may be reduced. These patients require close monitoring for evidence of bowel ischemia. Conversely, patients with strangulated hernias causing SBO may require bowel resection.

Nonviable bowel is resected; a decision is then made whether it is safe to perform a primary anastomosis or an ileostomy. If bowel viability is uncertain or equivocal, measures can be taken to assist in the decision of whether to resect. These include the use of intravascular fluorescein to visually demonstrate perfusion (ischemic bowel will not fluoresce), Doppler ultrasound assessment of mesenteric pulsations, or visual assessment of capillary filling of serosal blood vessels. If doubt still exists, the bowel may be kept in situ and a "2nd-look" laparotomy performed in 24 h.

Finally, Crohn's inflammation and/or stricture may require stricturoplasty if nonoperative management with bowel rest, decompression, and/or the addition of anti-inflammatory agents fails. Aggressive bowel resection in this group of patients should be avoided, as it may lead to malabsorption, diarrhea, fistula formation, or anastomotic leaks.

Conclusion

Small bowel obstructions require judicious management with a thorough history, physical, and focused diagnostic workup. Most of these patients will improve without surgery, but for those who do require an operation, prompt and directed evaluation is critical.

38
Ulcer Disease and *Helicobacter pylori*

Chetan Aher and Daniel J. Deziel

Introduction

Peptic ulcer disease (PUD) involves defects in the gastric or duodenal mucosa resulting from an imbalance of gastric acid production and mucosal protection. Once a major cause of hospitalization and upper gastrointestinal surgery, the treatment of PUD was transformed by the discovery of a causal association with *Helicobacter pylori* (*H. pylori*) in the 1990s. Other etiologic factors include the use of nonsteroidal anti-inflammatory drugs (NSAIDs), steroids, tobacco, cancer, physiologic stress from burns (Curling's ulcer), head injury (Cushing's ulcer), surgery, critical illness, and conditions associated with hypersecretion of gastrin. In the United States, hospitalizations for PUD declined by 30–40 % between 1993 and 2006. This decrease has been attributed to the availability of diagnostic and therapeutic endoscopy and to the improved efficacy of medical therapy. However, PUD remains a condition that may require surgical intervention. The indications for operation in PUD include bleeding, perforation, obstruction, intractability, and inability to exclude neoplasm.

Signs and Symptoms

Symptoms in patients with PUD can, but do not necessarily, differ based on the locations of the ulcer (Fig. 38.1). Patients with either gastric or duodenal ulcers usually present with epigastric abdominal pain that may be relieved by food or antacids; pain from a duodenal ulcer can radiate to the back. Gastric ulcers may be more symptomatic in the morning or in a fasting state when intragastric pH is lower. Duodenal ulcers may be aggravated by eating as the pH drops in the duodenum after gastric emptying. Hematemesis, melena, or bright red blood per rectum (BRBPR) are all possible manifestations of a bleeding ulcer. Gastric ulcers near the pylorus may present with nausea and vomiting from gastric outlet obstruction secondary to acute edema or chronic fibrosis. Special attention should be paid to other signs and symptoms such as weight loss, anemia, dysphagia, or a palpable abdominal mass, which may indicate a malignant process.

Physical Examination

Physical examination should focus on vital signs and a thorough abdominal exam. Patients with bleeding may show signs of anemia, shock, or hemoccult-positive stool. Patients with perforation may exhibit signs of peritonitis.

Workup

Diagnostic testing for suspected PUD depends on the manner in which the patient presents. Workup of the stable outpatient should initially focus on confirming the presence of *H. pylori* infection, which can be achieved via tissue urease test, urease breath test, or serologically via anti-*H. pylori* IgG. Patients found to be positive should undergo "triple therapy" with a proton pump inhibitor (PPI) and double antibiotic coverage. Though multiple regimens exist, most commonly a PPI in addition to amoxicillin and clarithromycin is used. Patients older than 45 years, those with associated gastroesophageal reflux disease, or those with signs and symptoms refractory to triple therapy should undergo esophagogastroduodenoscopy (EGD) for further evaluation. Patients with recurrent or refractory disease should undergo workup for gastrinoma by obtaining a fasting gastrin level while off all medical therapy for at least 2 weeks.

Treatment of the Uncomplicated Patient

Treatment of uncomplicated PUD includes pharmacologic acid reduction and eradication of *H. pylori*. In addition, patients are counseled on lifestyle changes to eliminate

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_38, © Springer New York 2015

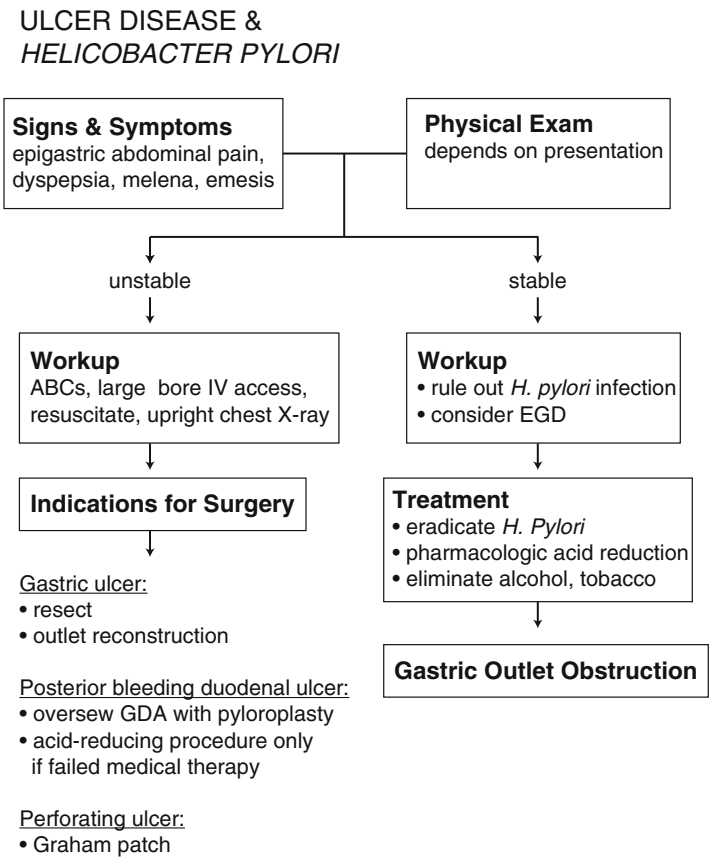

FIG. 38.1 Treatment algorithm for ulcer disease. *ABCs* airway, breathing, circulation, *IV* intravenous, *CXR* chest X-ray, *EGD* esophagogastroduodenoscopy, *GDA* gastroduodenal artery

offending substances such as alcohol, tobacco, and caffeine and to avoid common medications that are mucosal irritants (NSAIDS, aspirin). In the acute setting, a patient's vital signs direct the pace and order of workup. For the stable patient, routine labs including complete blood count (CBC) and chemistry panel should be obtained to assess leukocytosis, anemia, or electrolyte abnormalities. An upright chest X-ray should be obtained to assess for pneumoperitoneum. If the above workup is negative, and the history and physical does not include any signs or symptoms of bleeding, outpatient endoscopy may be pursued. Instability warrants prompt investigation with the addition of type and screen for possible transfusion.

Surgery for Complicated PUD

This category includes patients with significant bleeding, intractable pain, perforation, and obstruction.

Surgical indications for bleeding ulcers include failure of endoscopic hemostasis, ongoing hemodynamic instability despite aggressive resuscitation (greater than 6 units pRBC transfusion), recurrent hemorrhage after two endoscopic interventions on the same bleeding site, or an ongoing slower hemorrhage requiring repeated transfusions. Additional considerations for surgery include refusal of transfusion, inability to rule out malignancy, large ulcers, and obstruction.

Gastric ulcers meeting the aforementioned criteria require resection and gastric outlet reconstruction with options including Billroth I (gastroduodenostomy), Billroth II (gastrojejunostomy), or Roux-en-Y gastrojejunostomy.

Posterior duodenal ulcers meeting the aforementioned criteria require duodenotomy or pyloroduodenotomy to oversew the bleeding vessel, and the gastroduodenal artery (GDA) is suture ligated. Pyloroplasty is then undertaken for closure. If the patient failed appropriate medical acid suppression therapy, an acid-reducing procedure should be performed (truncal/selective/highly selective vagotomy). If the patient was untreated or undertreated, this portion of the procedure may be deferred in lieu of medical therapy.

Perforated duodenal ulcers, usually anterior, with resulting pneumoperitoneum require surgical intervention with omental patch closure. Patients who failed appropriate medical therapy should also have an acid reduction procedure as described previously. Ten to twenty percent of perforated duodenal ulcers have no evidence of free intraperitoneal air on upright chest X-ray. Computed tomography (CT) of the abdomen/pelvis with PO water-soluble contrast may demonstrate free air not detectable on plain radiography or free

fluid; however, given that some perforated ulcers will seal spontaneously, the perforation may not be detectable regardless of imaging modality. These patients are candidates for nonoperative management should they improve clinically.

While laparoscopic versions of all of the aforementioned procedures have been described, laparoscopic repair is most commonly performed on perforated duodenal ulcers since the omental (Graham) patch is the easiest to perform laparoscopically. A meta-analysis of laparoscopic versus open repair of perforations demonstrated that laparoscopic repair is safe and feasible and may be associated with less blood, quicker recovery, a lower rate of wound infection, and decreased mortality.

Gastric Outlet Obstruction

Gastric outlet obstruction can occur acutely due to edema or as a late complication resulting from chronic disease with fibrotic stricture. Once the most common cause of gastric outlet obstruction, the incidence decreased significantly after the advent of PPIs. At present, malignancy is the most common cause of gastric outlet obstruction. Obstruction is usually due to a combination of edema, inflammation, and spasm, which can progress to fibrosis and scarring and ultimately gastric atony from long-term obstruction. Patients present with nausea, vomiting, weight loss, early satiety, bloating, and epigastric pain. Both medical and surgical therapies exist, and medical treatment should be attempted initially. In the acute setting, fluid and electrolyte status should be optimized. All NSAIDs should be stopped, and intravenous PPI should be started with progression to liquid formulations as the patient begins to pass liquids. If medical therapy alone is insufficient or ineffective, endoscopy with biopsy should be undertaken, followed by endoscopic balloon dilation. Medical therapy may be continued throughout this process. Patients refractory to medical or endoscopic therapy require surgery, with reconstructive options as discussed previously.

39
Ischemic Bowel

Gregory Stettler

Introduction

Mesenteric ischemia is an uncommon problem caused by a reduction of blood flow in the mesenteric vasculature secondary to occlusion, vasospasm, and/or hypoperfusion. If not rapidly identified, mesenteric ischemia can lead to catastrophic clinical consequences including bowel infarction, sepsis, and death (Fig. 39.1). The elderly are more likely to be affected, but mesenteric ischemia may affect any age group. The clinical presentation of ischemic bowel depends on the underlying pathologic abnormalities as well as other comorbid medical conditions. Classically, patients will present with *abdominal pain that is out of proportion to the physical exam findings*. Furthermore, the *pain is often unresponsive to narcotic analgesia*. Dehydration and third spacing of fluids may lead to changes in mental status, tachycardia, tachypnea, and in severe cases, circulatory collapse.

Laboratory findings include anion gap metabolic acidosis, elevated lactate levels, leukocytosis, and hemoconcentration. Once the diagnosis of mesenteric ischemia is made, treatment should be initiated immediately. Delayed treatment will decrease the chance for successful bowel salvage and increase risk of death. Management in an intensive care setting is mandated in a patient suspected of having mesenteric ischemia.

The causes of ischemic bowel include acute mesenteric arterial occlusion from an emboli or thrombosis, nonocclusive mesenteric insufficiency, mesenteric venous occlusion, and chronic mesenteric insufficiency.

Acute Mesenteric Arterial Occlusion

Acute mesenteric arterial occlusion is often caused by an embolus that occludes the superior mesenteric artery (SMA). The celiac or inferior mesenteric artery (IMA) may also be affected. The SMA is most often affected because of its large caliber and narrow branching angle from the abdominal aorta. Acute mesenteric ischemia may be due to acute plaque changes at the origin of the SMA. Most mesenteric emboli have a cardiac origin. Myocardial ischemia or infarction, atrial fibrillation or other tachyarrhythmias, endocarditis, cardiomyopathies, ventricular aneurysms, and valvular disorders are risk factors for the development of a cardiac thrombus. Acute mesenteric ischemia should be considered in any patient with a history of cardiac disease or previous embolic events who presents with acute onset, severe abdominal pain accompanied by diarrhea that may be bloody. As ischemia progresses to the point of full-thickness necrosis, pain will worsen and can be followed by bowel perforation and peritonitis.

Radiographic findings are often nonspecific but may show edematous, fluid-filled bowel loops. Angiography, duplex ultrasound, CT, or MRA may be used to make a diagnosis; angiography remains the diagnostic gold standard and often reveals occlusion of one or more of the major splanchnic arteries. Duplex ultrasound is another diagnostic tool, but may be limited by body habitus, overlying bowel gas patterns, or operator experience. Multidetector helical CT allows for visualization of stenotic regions of the mesenteric vasculature. MRA is limited by its potential inability to visualize the IMA.

Once the diagnosis is made, treatment should be initiated without delay. This includes active resuscitation as well as addressing the underlying problem that led to the embolus. If there are no signs of peritonitis and angiographic evidence of mesenteric occlusion is present, an intra-arterial infusion of plasminogen activator (TPA) may be used to dissolve the arterial occlusion. Systemic heparinization is also indicated at this time. Ineffective lysis of the occlusion or the development of peritonitis is an indication for surgical intervention. Infusions of Papaverine or other vasodilators can be administered through the angiographic catheter. For acute mesenteric embolism, a standard embolectomy via transverse arteriotomy may be performed. If the acute arterial occlusion originates from arterial thrombosis due to atherosclerotic disease, a vascular bypass graft is usually necessary. In this case, the bypass graft can originate from the supraceliac or infrarenal aorta.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_39, © Springer New York 2015

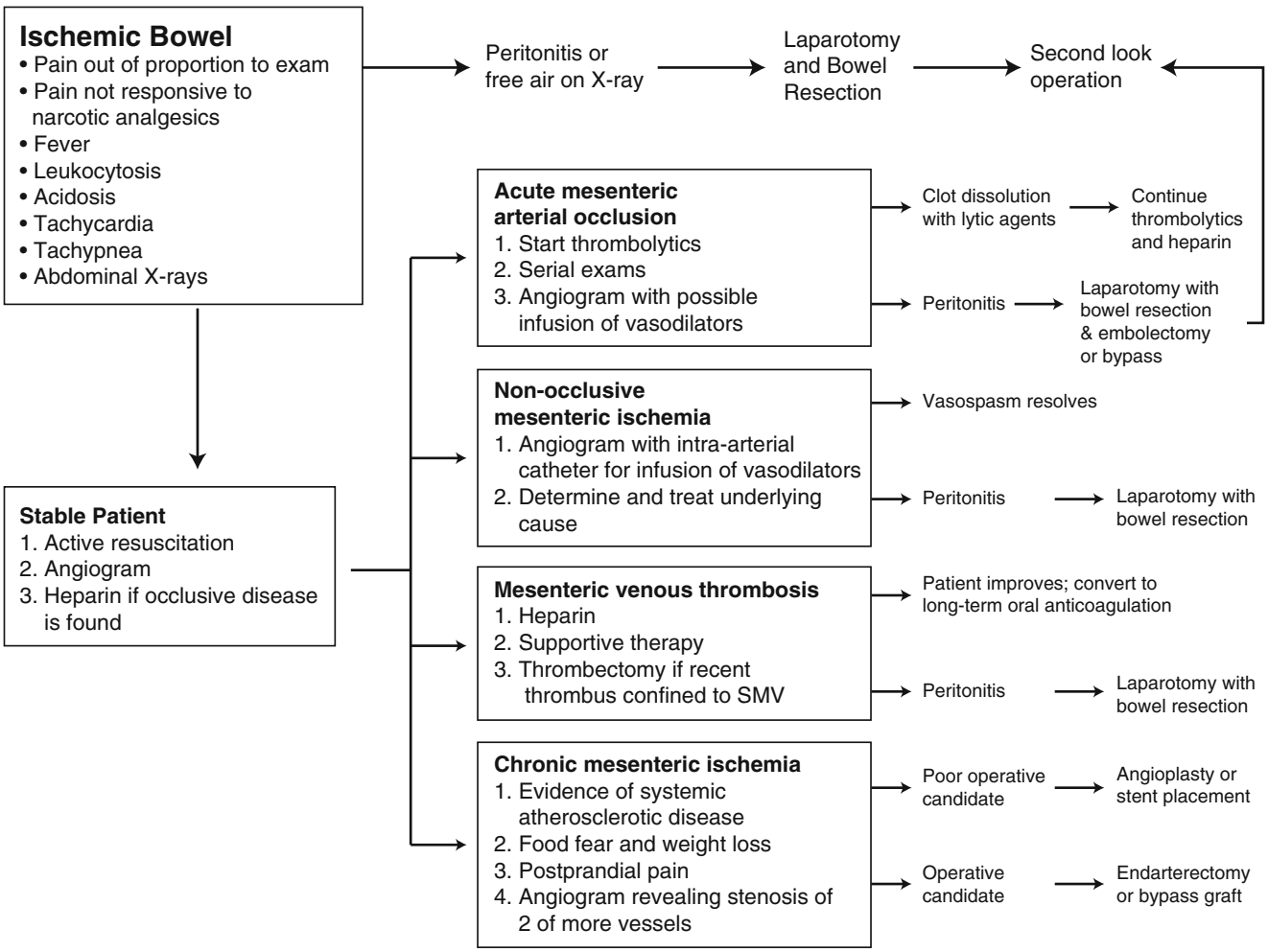

FIG. 39.1 Ischemic bowel treatment algorithm. *SMV* superior mesenteric vein

In all cases of acute mesenteric ischemia, blood flow should be reestablished followed by resection of necrotic bowel. There are various methods of determining bowel viability intraoperatively. The remaining bowel can be assessed for color, arterial pulsations, and capillary refill. Doppler ultrasound can also be used to determine blood flow. Examination of the bowel with a Wood lamp following administration of 1 g of intravenous fluorescein can help distinguish poorly perfused from viable bowel. Finally, laser Doppler flometry (LDF) and visible light spectrophotometry (VLS) can assess bowel viability.

Following an initial laparotomy with revascularization and resection of necrotic bowel, a "second-look" operation is indicated 24–48 h later to reassess the viability of the remaining bowel. At this point, the decision to restore intestinal continuity will be based on the overall condition of the patient, the presence of intraperitoneal contamination, and the state of the bowel perfusion following revascularization. Permanent or temporary stomas may be created as well. Operative mortality in patients undergoing revascularization can range from 25 to 90 % and depends greatly on timely diagnosis and intervention. Some patients will survive massive bowel resection and develop short-gut syndrome, requiring long-term parenteral nutritional supplementation or eventual bowel transplantation.

Nonocclusive Mesenteric Insufficiency

Nonocclusive mesenteric insufficiency (NOMI) results from intermittent severe vasoconstriction of the splanchnic blood supply caused by a low cardiac output state. Splanchnic vasoconstriction is a physiologic response to hypovolemic and cardiogenic shock. The use of vasopressors to maintain hemodynamic stability in critically ill patients will cause further mesenteric vasoconstriction and increase the risk of developing NOMI. Risk factors for developing NOMI include age older than 50 years, myocardial infarction or congestive heart failure, aortic valve insufficiency, cardiopulmonary bypass procedures, renal or hepatic disease, and

major abdominal or cardiovascular surgery. Similar to acute mesenteric arterial occlusions, patients with NOMI will present with sudden abdominal pain that is out of proportion to physical exam findings. Angiogram of the mesenteric vasculature is the primary diagnostic method and has the added benefit of being simultaneously therapeutic. The classic finding of NOMI on angiography is large vessel "beading" of mesenteric branches from focal vasospasm. Vasoconstriction of smaller mesenteric branches may produce a "pruned tree" appearance on angiography. Once diagnosis is made, therapy includes infusion of Papaverine through the angiographic catheter. The ultimate treatment for NOMI is to treat the underlying cause of splanchnic hypoperfusion. Peritoneal signs on physical exam are suggestive of advanced pathology and warrant immediate surgical intervention. Nonviable or necrotic bowel is resected as needed. The methods employed to determine bowel viability during surgery are identical to those discussed previously. Given that many patients with NOMI have severe underlying disease that triggers the splanchnic vasospasm, mortality rates are high.

Mesenteric Venous Thrombosis

Mesenteric venous thrombosis results in bowel ischemia due to reduced venous drainage of the mesenteric vasculature. It is the least common cause of mesenteric ischemia. Historically, most cases were thought to be due to intra-abdominal pathologic conditions such as malignancy, intra-abdominal sepsis, and pancreatitis or were classified as idiopathic. However, with improved diagnostic techniques, more cases have been shown to be associated with primary clotting disorders.

Intestinal ischemia following mesenteric venous thrombosis is due to the resistance in mesenteric venous blood flow. This leads to bowel wall edema and ultimately systemic hypotension. As a result, arterial blood flow decreases. Patients will often present with vague, insidious abdominal pain. Due to possible bowel infarction, stool samples may be hemoccult positive. As with other types of bowel ischemia, angiography is the diagnostic gold standard and will confirm the presence of venous congestion.

Once a diagnosis is made, current treatment protocols dictate systemic heparinization and eventual long-term anticoagulation with Coumadin. In the absence of ongoing thrombotic disorders, anticoagulation may be limited from 6 to 12 months. Surgical exploration is not necessary in all patients with mesenteric venous thrombosis. Treatment primarily involves supportive management unless there is progression to bowel infarction or peritonitis, which mandates immediate surgery. Thrombectomy may be a therapeutic possibility when the thrombus is recent and is confined to the superior mesenteric vein (SMV). More diffuse types of venous thrombosis will preclude the use of surgical thrombectomy. Mortality ultimately depends on whether the venous thrombosis is acute or chronic. Long-term survival is 30–40 % in patients with acute venous thrombosis and 80 % in patients with chronic venous thrombosis.

Chronic Mesenteric Ischemia

Chronic mesenteric ischemia is episodic or constant intestinal hypoperfusion of the mesenteric vasculature. The most common cause is atherosclerosis of the proximal portions of the celiac artery, SMA, or IMA. Factors that increase the risk of atherosclerotic disease in the heart or carotid arteries will also affect the mesenteric vasculature. These risk factors include smoking hypertension, diabetes mellitus, and hypercholesterolemia.

Other less common etiologies may include dissection, vasculitis, or cocaine abuse. Typically, two of the three major splanchnic vessels must be occluded before symptoms occur due to collateralization between vessels. After eating, the increased need for mesenteric blood flow, coupled with the presence of a fixed, occlusive plaque within the splanchnic vasculature, prevents adequate circulation to the mesenteric vessels. Classic symptoms of chronic mesenteric ischemia include postprandial abdominal pain associated with significant weight loss, food fear, nausea, vomiting, and/or diarrhea. Due to the fact that pain associated with chronic mesenteric insufficiency is most commonly postprandial, this disease is often referred to as "intestinal angina."

Physical exam findings will include weight loss, malnutrition, evidence of systemic atherosclerotic disease, and an abdominal bruit in approximately 50 % of patients. Diagnosis of chronic mesenteric ischemia is similar to that of acute mesenteric arterial occlusion, with the gold standard being angiography. Definitive treatment of chronic mesenteric ischemia occurs through either open or endovascular surgical techniques. Open surgical techniques include transaortic endarterectomy of the occluding plaque, direct reimplantation on the aorta, or bypass grafting using either prosthetic or autogenous conduits to create new paths for blood to flow. An alternative to open surgery is endovascular repair, which includes angioplasty or stent placement of stenotic lesions within the mesenteric vasculature. Lower morbidity and mortality associated with endovascular revascularization have led to increased popularity of this technique as a first-line treatment for patients with chronic mesenteric ischemia.

40
Crohn's Disease

Maria C. Mora Pinzon and Dana M. Hayden

General Considerations

Crohn's disease (CD) is a chronic inflammatory disease affecting approximately 500,000 individuals in the USA. It is characterized by focal, asymmetric, transmural inflammation that may involve any part of the gastrointestinal tract (Fig. 40.1). The disease is more frequent in females and has a bimodal distribution; it is most frequently diagnosed between 15 and 35 years old with a secondary peak between ages 60 and 80. Its pathogenesis is not well understood, but it appears to be multifactorial; recent data suggest that CD involves a genetic predisposition and dysregulation of the inflammatory response and may be triggered by environmental factors (smoking, oral contraceptives, and anti-inflammatory drugs). The disease is more common in urban areas in Europe, the UK and the USA, with a predisposition among Jewish populations; the prevalence is lower in non-Jewish white, African American, Hispanic, and Asian populations.

Manifestations

CD has been classified according to location: small bowel only (25 % of patients), small bowel and colon (50 %), colon only (20 %), and perianal. Uncommon locations include the esophagus, stomach, and duodenum. The type of disease may be described as inflammatory, fibrostenotic, and fistulizing or by severity (mild, moderate, severe). Presentation will depend on the segment of the intestinal tract affected. CD initiates insidiously; however, on rare occasions, the disease may present in a fulminant manner, such as toxic colitis. Most patients have relapsing episodes followed by remission.

The most common symptoms include: chronic or nocturnal watery diarrhea, colicky abdominal pain relieved by defecation, fever, and weight loss. Rectal bleeding may be present. Physical examination may reveal abdominal distension, a mass, tenderness, or perianal disease (abscesses or fistulas). Extraintestinal manifestations (anterior uveitis, erythema nodosum, nephrolithiasis, osteoporosis, spondyloarthritis) are present in almost 25 % of patients. Patients with ileitis may present with pain in the right lower quadrant, which mimics acute appendicitis.

Diagnosis

Patients with the aforementioned clinical manifestations should be evaluated for inflammatory and infectious bowel disorders, including: celiac disease, diverticulitis, infections (*Yersinia*, *Mycobacterium*, *Campylobacter*), ulcerative colitis (UC), CD, irritable bowel syndrome, and food intolerance. The diagnosis of Crohn's is established by pathologic, radiographic, endoscopic, and laboratory means. The gold standard for diagnosis is colonoscopy with intubation and evaluation of the terminal ileum.

- *Laboratory*: Laboratory studies include: complete blood count with differential, stool culture for ova and parasites, testing for *Clostridium difficile* toxin, C-reactive protein, erythrocyte sedimentation rate, antibodies to *Escherichia coli* outer membrane, antibodies to *Saccharomyces cerevisiae (ASCA),* and perineural antineutrophil cytoplasmatic antibodies (pANCA). A positive pANCA is suggestive of ulcerative colitis, while positive ASCA is more likely to be positive in Crohn's. Further evaluation of nutritional status of the patient should be performed (albumin, vitamin B12, vitamin D, folate).
- *Imaging*: Imaging studies include: computed axial tomography (CT), magnetic resonance imaging (MRI), and abdominal ultrasound. These studies may show evidence of inflammation and complications of CD, such as abscess, fistula, obstruction, or perforation. Double-contrast enema (barium or Gastrografin) is used to delineate the extent of colonic disease and is more sensitive than CT in the detection of fistulas involving the colon. Fluoroscopy with small bowel follow-through will identify strictures, fistulas, and obstruction. Newer modalities include CT and

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_40, © Springer New York 2015

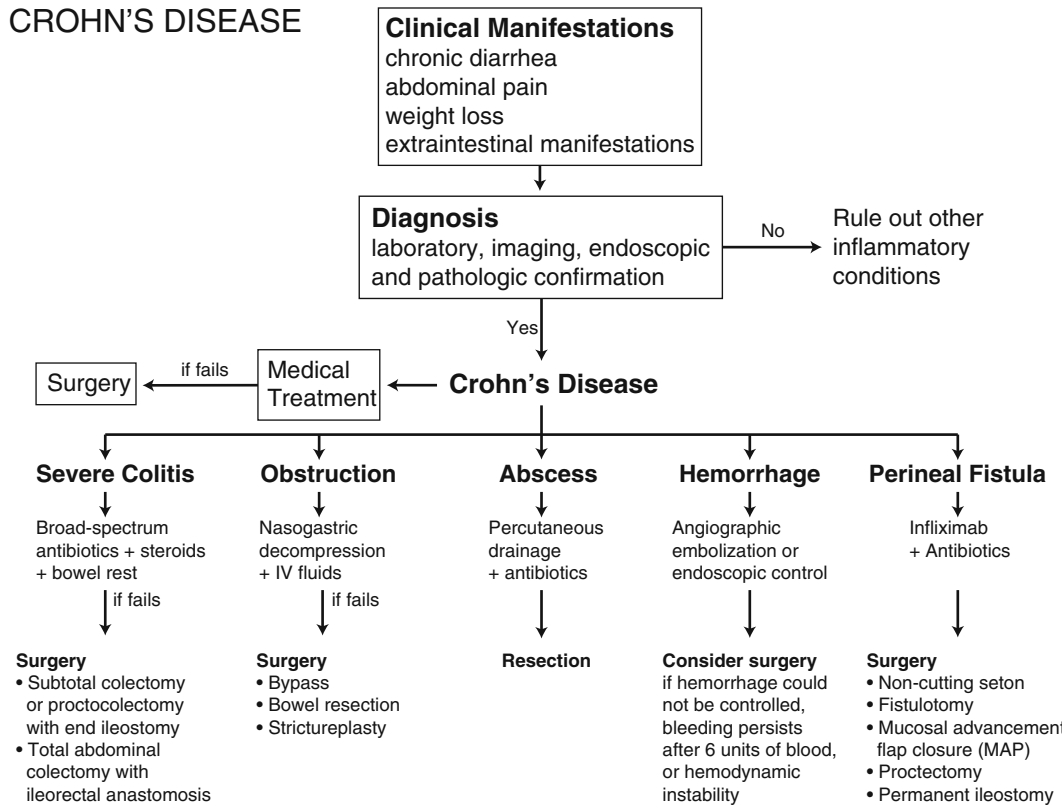

FIG. 40.1 Treatment algorithm for Crohn's disease. *IV* intravenous

MRI enterography, which may identify active inflammation throughout the intestinal tract. Capsule endoscopy allows for endoluminal visualization of the small bowel, but it should be avoided in patients with strictures due to the risk of capsule retention. MRI is also used to evaluate perianal Crohn's disease helping to identify perirectal, perineal, and rectovaginal fistulas and abscesses.

- *Endoscopy (upper and lower, enteroscopy):* findings include ulcerations, strictures, skip lesions, "cobblestoning." Pathology may show non-caseating granulomas and transmural inflammation.

Medical Treatment

The goal of the medical treatment of CD is: (1) to control acute exacerbations and symptoms and (2) to maintain remission with minimal adverse effects. There are several categories of medications including: mesalamine products (sulfasalazine and 5-aminosalicylic acid), antibiotics (metronidazole, fluoroquinolones), corticosteroid therapy, azathioprine, 6-mercaptopurine, methotrexate, and antitumor necrosis factor (anti-TNF) agents, infliximab (Remicade), adalimumab (Humira), and certolizumab pegol (Cimzia). Two theories of medical management include the "step-up" (initiated with 5-ASA and advances to corticosteroids and immunomodulators) or "step-down" practice (for moderate-severe disease, initiated with immunomodulators and anti-TNF medications). The selection of medication type, dose, formulation, and combination depends on disease location and severity, adverse effects of the medication, and patient tolerance and clinical status.

Surgery

Approximately 65 % of patients with Crohn's disease will require surgery at some point in their lifetime. Indications for surgical intervention include: failure of medical treatment, intolerance of medication side effects, complications of disease (obstruction, fistula, perforation, hemorrhage, abscess), or the development of dysplasia or cancer. Choice of operation depends on the location and type of disease (fistulizing, stricturing). The chief objective of surgery is to correct the abnormality yet conserve as much bowel as possible.

For small bowel strictures, resection and strictureplasty are the most commonly performed operations. The choice depends on the extent of disease, number and location of strictures present, and the patient's history of previous small bowel resections. Again, conservation of absorption surface is paramount. Resection can be performed if there are multiple strictures within a short segment of bowel or an

isolated stricture in a patient with no previous resections. Studies have shown that there is no difference between stapled and hand-sewn anastomosis with respect to Crohn's recurrence; however, large side-to-side anastomoses have lower recurrence rates. Resection should be performed conservatively to grossly normal-appearing margins; there is no merit in obtaining histologic confirmation of negative margins. Although technically difficult, laparoscopy has proven to be safe and effective in Crohn's disease, reducing length of hospitalization and recovery period. Primary anastomosis should be avoided in patients with severe malnutrition or sepsis.

Endoscopic recurrence after resection occurs in about 80 % of CD patients, more frequently at the anastomotic site or just proximal to it; 30 % of patients will require further surgery. Postoperative medical management will decrease recurrence after surgery, especially among patients at higher risk (active tobacco smoking and severe disease).

Small bowel strictures can also be treated with strictureplasty. If the patient had previous major small bowel resections, has multiple strictures diffusely throughout the small intestine, or has a risk of short bowel syndrome, strictureplasty is preferred. Heinecke-Mikulicz strictureplasty is performed for short strictures or Finney for longer strictures over 10 cm. There has been some concern for the risk of cancer within strictures; therefore, some surgeons will biopsy the stricture before completion of the strictureplasty. If the disease is very proximal or the inflammation makes resection too difficult, intestinal bypass or proximal diversion can be performed.

If the patient presents acutely, intravenous hydration, bowel rest and antibiotics should be started immediately. If peritonitis and perforation are present, surgery should not be delayed. Abscess drainage should be performed percutaneously if possible. Obstruction should be treated with bowel decompression, intravenous hydration, antibiotics, and steroids; nonresponse requires surgery. The time at which to intervene will vary from one surgeon to the next, but being conservative is best for the patient as long as he/she is clinically stable. If the patient presents with severe colitis (defined as six or more bloody bowel movements per day and evidence of systemic toxicity), immediate treatment should include bowel rest, broad-spectrum antibiotics, and intravenous steroids. If the patient's condition deteriorates or fails to improve within 24–48 h, options include subtotal colectomy with end ileostomy or initiation of treatment with biologic therapy.

Management of Crohn's-related hemorrhage will depend on the location and severity. The source of bleeding may be identified and controlled with endoscopy or mesenteric angiography. Surgery should be considered if hemorrhage cannot be controlled with endoscopy or interventional radiology, bleeding persists after 6 units of blood have been transfused, the bleeding recurs within a short period of time, or if the patient is hemodynamically unstable.

Treatment of perianal Crohn's disease is with a combination of medical and surgical means. Symptomatic perianal fistulas are initially treated with antibiotics (metronidazole and fluoroquinolones) and anti-TNF medications. Surgery is used to control sepsis and prevent symptoms such as pain from recurrent abscesses. Low-lying simple fistulas can be treated with fistulotomy, while complex fistulas are treated with long-term draining setons. It is important to control active perianal Crohn's and proctitis before attempting to surgically manage fistulas due to the high failure rate. Recent studies describe use of non-cutting setons in association with infliximab as safe and effective, with healing rates of 24–75 %. Patients with refractory fistulas may require advancement flap closure, proctectomy, or permanent diversion to control perianal sepsis.

Conclusion

In conclusion, abscesses are promptly drained to obtain immediate relief; fistulas are approached with a combination of medical and surgical means. Active Crohn's is controlled medically; simple fistulas may be then treated with fistulotomy. Complex fistulas (through the sphincter) are treated with non-cutting setons, maximizing medical therapy with biologic medications, while reserving flap procedures and/or proctectomy for persistent, refractory, symptomatic disease.

41
Ulcerative Colitis

Daniel Rinewalt and Dana M. Hayden

Introduction

Ulcerative colitis (UC) is an idiopathic inflammatory bowel disease more common in developed countries and most often presenting between ages 15 and 30 with an additional peak in the sixth decade. Many possible etiologies have been implicated, including genetic mutations, environmental factors, infectious agents, and immunologic disturbances; however, no cause has been conclusively determined. The development of ulcerative colitis appears to be multifactorial; genetic markers have been identified, the disease runs in families, and it also responds to immunosuppressive medications (Fig. 41.1). Involvement of the rectum with continuous distribution of inflammation proximally, not involving the small intestine, is the classic feature that distinguishes UC from other inflammatory bowel conditions such as Crohn's disease (CD). However, it is important to note that this presentation may vary, making the diagnosis difficult in certain cases.

Beside symptoms of UC affecting quality of life, this condition has also been found to be a strong, independent risk factor for colon and rectal cancer as well as primary sclerosing cholangitis (with subsequent cirrhosis and bile duct cancer); UC requires close surveillance to avoid these potentially fatal complications.

Clinical Presentation

Signs and symptoms of ulcerative colitis can be divided into gastrointestinal (most common) or extraintestinal. The typical clinical presentation includes crampy abdominal pain, urgency, and watery or bloody diarrhea mixed with mucus. Other symptoms include anemia, weight loss, malnutrition, and failure to thrive resulting from long-standing, severe disease. A variety of extraintestinal symptoms may be present, including but not limited to: aphthous ulcers, uveitis, arthritis, erythema nodosum, pyoderma gangrenosum, deep vein thrombosis, and pericarditis. Presentation may vary from a mild insidious course with only diarrhea and minimal abdominal pain to a more sudden onset of explosive bloody diarrhea, severe abdominal pain with fever, tachycardia, and dehydration. A small subset of patients will present with fulminant toxic colitis that may or may not involve significant colonic dilatation but increases risk of bowel perforation, sepsis, acidosis, and shock. This presentation, while rare, can be potentially fatal and requires immediate recognition and treatment.

Diagnosis

No specific laboratory tests can diagnose ulcerative colitis, and physical exam is usually nonspecific. Endoscopy remains the definitive diagnostic modality for UC and other inflammatory bowel diseases. Flexible sigmoidoscopy with biopsy is usually sufficient for diagnosis; however, a complete colonoscopy may be required in cases where uncertainty remains, allowing for evaluation of the terminal ileum. Utilizing colonoscopy in the setting of acute inflammation, however, increases the risk of perforation significantly and is contraindicated when the presentation is severe. Typical findings on endoscopy include friable mucosa, exudates, pseudopolyps, superficial ulcers, and diffuse erythema. The rectum is invariably involved, with potential continuous spread throughout the entire colon.

Histologic examination will reveal inflammation of the mucosa and submucosa, crypt abscesses, extensive infiltration of inflammatory cells, mucin depletion, and disturbed crypt architecture. Antineutrophil cytoplasmic antibody (pANCA) serum marker may be positive.

Other diagnostic methods include plain radiographs to rule out perforation in the acutely ill patient with severe abdominal pain. Computed tomography (CT) scan or barium enema may reveal shortening of the colon and loss of haustral markings; however, these findings are not sufficient for definitive diagnosis and cannot differentiate between other types

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_41, © Springer New York 2015

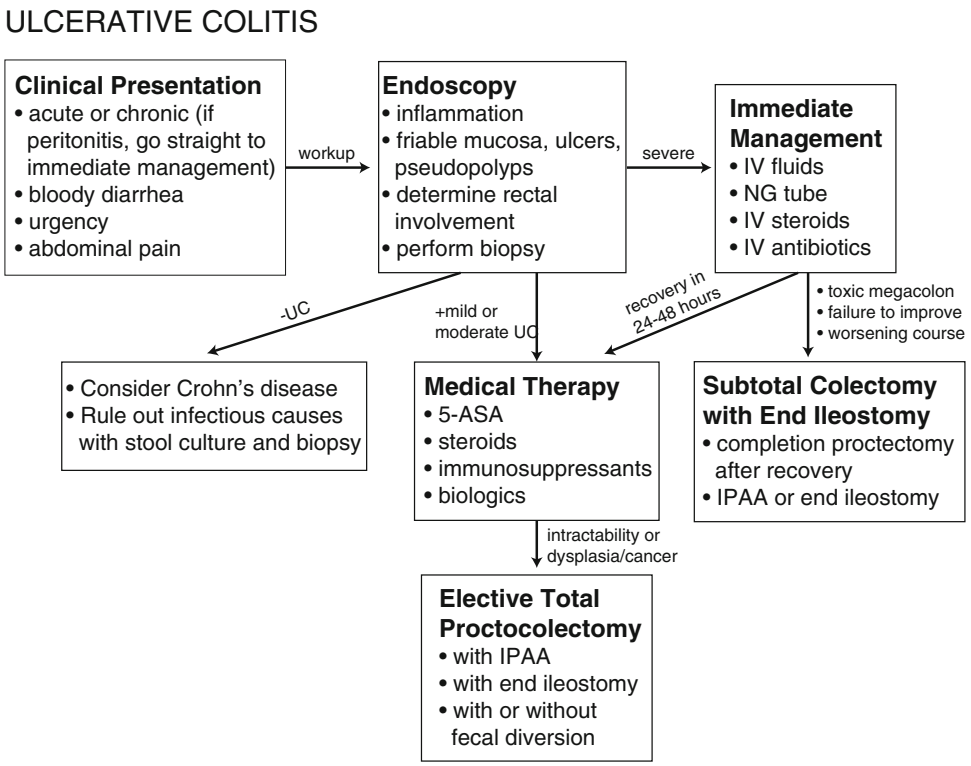

FIG. 41.1 Treatment algorithm for ulcerative colitis. *UC* ulcerative colitis, *IPAA* ileal pouch-anal anastomosis, *IV* intravenous, *NG* nasogastric, *5-ASA* 5-aminosalicylic acid

of inflammatory or infectious diseases. In order to rule out infectious causes of colitis, stool cultures should be routinely sent in addition to blood cultures in a severely ill patient.

Treatment

Initial treatment in most cases of ulcerative colitis begins with medical therapy in order to relieve symptoms and induce remission. For mild symptoms, such as watery diarrhea, treatment with medications to slow intestinal transit, such as loperamide or bulking agents, may be sufficient.

For more severe presentations, aminosalicylates, steroids, and other immunosuppressants are the mainstay of treatment. Aminosalicylates (5-ASA, delayed release, oral, suppositories, enemas) are common first-line medications for moderate acute UC as well as maintenance therapy for long-term remission. Steroid therapy is typically used for the acute severe disease, but should not be used for long-term therapy secondary to serious, well-described side effects. Immunosuppressants including azathioprine, 6-mercaptopurine, and methotrexate are most effective for long-term maintenance therapy and not for acute presentation since their treatment effects may take months to manifest. More recently, biologics (antitumor necrosis factor medications) have also been shown to treat acute and severe UC and effectively maintain remission as well.

Indications for surgery in UC include:

1. Failure or noncompliance with medical therapy
2. Inability to tolerate medication side effects
3. Massive or persistent colonic bleeding not amenable to less invasive therapy (endoscopy or embolization)
4. Fulminant colitis or toxic megacolon
5. Chronic anemia, malnutrition, failure to thrive
6. Perforation
7. Obstruction
8. Dysplasia or cancer

One advantage of surgical treatment in UC is that, unlike Crohn's, complete resection of the colon and rectum is curative. Elective surgical options include total proctocolectomy with end (permanent) ileostomy or total proctocolectomy with ileal pouch-anal anastomosis (IPAA). Both options have the advantage of removing almost all potential tissue involved in the disease process and therefore eliminating almost all risk of disease recurrence or progression to colon cancer (small amount of distal rectum remains unless mucosectomy performed or anal canal removed). The main drawback of end ileostomy is the requirement of permanent ileostomy; however, this may be necessary in patients with poor sphincter function (previous surgery, trauma, elderly patients). Total proctocolectomy with IPAA preserves the anal sphincters and creates a reservoir (the ileal pouch) that allows the patient to defecate through the

anus with relatively normal bowel function. This operation is frequently performed in two stages and can be performed laparoscopically. The first stage involves removal of the colon and rectum, formation of the ileal pouch anal anastomosis, and creation of the diverting ileostomy. The second is reversal of the loop ileostomy after the IPAA heals and the patient recovers. Alternatively, few colorectal surgeons question the need for fecal diversion and perform a one-stage procedure.

Patients presenting with fulminant colitis require immediate attention. Aggressive intravenous fluid resuscitation, nasogastric decompression, and broad-spectrum intravenous antibiotics should begin without delay. Usually, medical treatment is initiated with high-dose intravenous steroids and a trial of conservative therapy with bowel rest and possible total parental nutrition. The patient should be closely monitored for 24–48 h. If the patient's condition worsens or fails to improve during this time, surgical intervention is indicated. In these circumstances when patients are critically ill, a subtotal colectomy (colectomy without proctectomy) is the procedure of choice due to its shorter operative time and less operative morbidity. Subtotal colectomy leaves the option for future sphincter preservation and pouch creation. In patients with severe UC, malnutrition, or anemia or colonic bleeding with hemodynamic instability or persistent bleeding with failure of nonsurgical management, a subtotal colectomy is recommended. After an appropriate recovery, completion proctocolectomy can be done with IPAA or end ileostomy.

42
Acute Appendicitis

Tasha M. Hughes and Minh B. Luu

Introduction

Reginald Fitz first described appendicitis in 1886, although the first appendectomy is thought to have been performed in 1735 by Claudius Amyand during an inguinal hernia repair. Appendectomy as we know it today was first performed by Willard Packard in 1867. While the presentation has not changed significantly the workup, surgical approach and morbidity and mortality from the disease have evolved significantly. *Appendicitis remains the most common cause of an acute abdomen* with 250,000 appendectomies performed annually in the United States. Appendicitis most frequently affects younger patients and is slightly more common in men than in women. The appendix is a blind-ended structure extending off the bowel lumen at the transition from the small to large intestine. *The pathophysiology of appendicitis is luminal obstruction*; however, the cause of obstruction varies. In the pediatric population, lymphoid hyperplasia is a common cause of appendiceal occlusion. In adults, fecal inspissation, plant seeds, or tumor may all cause obstruction, with the former comprising the vast majority of cases. Luminal obstruction leads to edema of the bowel wall, leading to venous congestion, which in turn generates further edema. Eventually, arterial insufficiency, ischemia, necrosis, and perforation will follow.

History and Physical Exam

Appendicitis is still thought by many to be a largely clinical diagnosis (Fig. 42.1). Pain is described as first appearing in the periumbilical location with subsequent migration to the right lower quadrant. This migrating pattern is attributed to stimulation first of T-10 level pain fibers (at the level of the umbilicus) followed by localized irritation of the peritoneum in the right lower quadrant (RLQ) once the inflammatory process has progressed. Pain is followed by nausea and vomiting several hours after initial onset. The pain may start abruptly and get progressively worse over a matter of hours to days. At the time of presentation, pain has usually been present for 24 h or less. Patients may report anorexia and fever or chills. Elderly patients may be present atypically and may even present with signs of sepsis. Pediatric patients may report a recent viral or bacterial illness preceding the RLQ pain. Pain with palpation over McBurney's point (one-third the distance between the umbilicus and anterior superior iliac spine) is classic in the diagnosis of appendicitis. Rovsing's sign is pain at McBurney's point with palpation of the left lower quadrant (LLQ) and suggests the presence of peritonitis. The psoas sign is pain with extension of the right hip. The obturator sign is pain with internal rotation of the right hip. Physical exam should also include a rectal exam and a pelvic exam. Pain with either may signal the presence of a pelvic abscess. The pelvic exam has the added benefit of ruling in or out pelvic inflammatory disease.

Diagnosis

Once appendicitis is suspected on clinical exam, it is the job of the clinician to determine if perforation is likely to have occurred. Involuntary guarding, rebound tenderness, and other signs of generalized peritonitis are suggestive of perforation. Additionally, patients who have had pain for greater than 24–36 h warrant increased suspicion for perforated appendicitis. After physical exam, basic lab tests should include WBC count, urinalysis, basic testing for STI in females, and beta-hCG in all females of childbearing potential. In many patients, particularly young males and children, with multiple positive elements of the history, physical exam, and laboratory abnormalities described being present, appendectomy may be justified without confirmation with imaging studies. Appendicitis in female patients and the elderly is often more difficult to diagnose on history and physical alone; therefore, imaging studies are usually performed.

Clinical scoring systems have been developed and well-studied. The modified Alvarado criteria consist of migratory

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_42, © Springer New York 2015

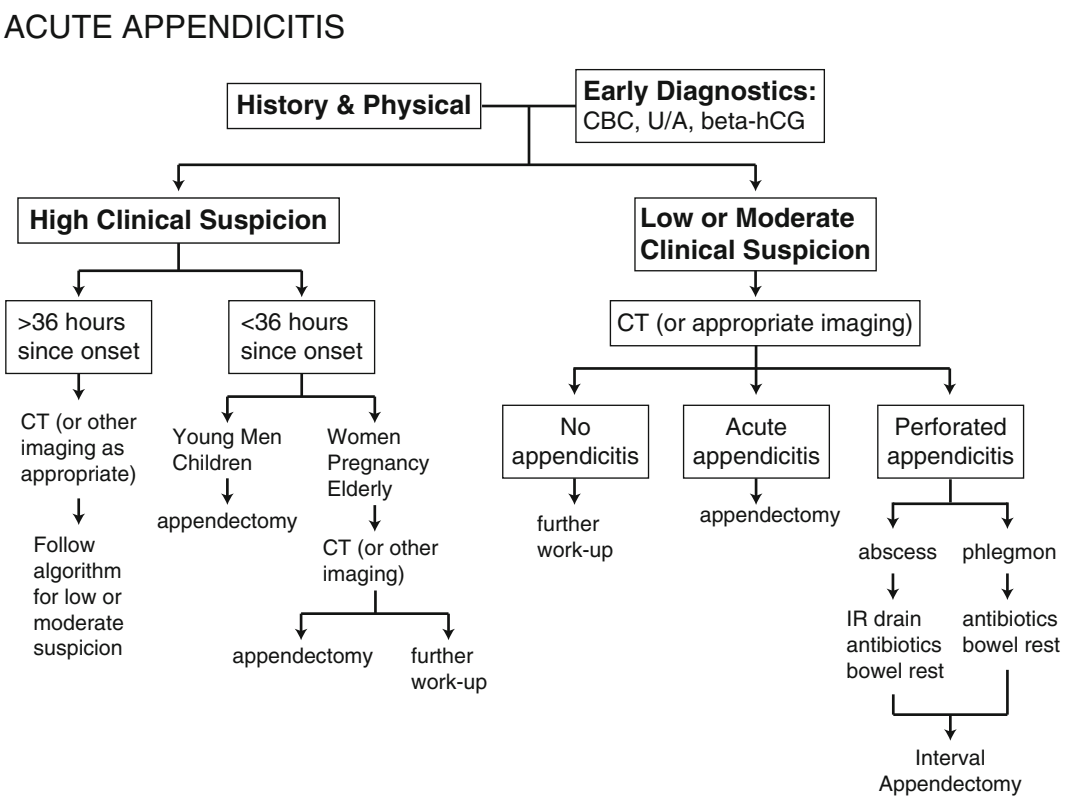

FIG. 42.1 Algorithm for treating acute appendicitis. *CBC* complete blood count, *U/A* urinalysis, *B-hCG* beta-hCG, *CT* computed tomography, *IR* interventional radiology

right iliac fossa pain (1 point), anorexia (1 point), nausea/vomiting (1 point), tenderness in the right iliac fossa (2 points), rebound tenderness in right iliac fossa (1 point), fever greater than 37.5 °C (1 point), and leukocytosis (2 points). Zero to three points corresponds to a low likelihood of appendicitis, four to six carries an intermediate likelihood, and a score of 7 or greater is associated with high probability of underlying appendicitis. Overall, the accuracy of the Alvarado score in diagnosing appendicitis is 83 % and the sensitivity of a score of 7 or greater is 95 %. However, this clinical scoring system is of limited use in females and has only been validated in young men and children. It is in female patients, elderly patients, and the overweight that addition of computed tomography (CT) scan provides the most benefit in diagnosis.

CT findings suggestive of appendicitis include dilation of the appendix to >6 mm, appendiceal wall thickening (>2 mm), periappendiceal fat stranding, appendiceal wall enhancement, and visualization of an appendicolith. Recent studies have suggested that in patients with an equivocal clinical presentation (Alvarado score 4–6), CT scan can improve diagnostic capacity and can help clinicians make more informed decisions on whether or not to proceed to the operating room. Large, multicenter trials have also demonstrated that over the time course of increasing CT utilization, the negative appendectomy rate (NAR) has decreased significantly. This decrease in NAR has been replicated in multiple studies and, in some, is thought to be even more significant when considering solely female patients or looking at obese patients in independent subgroup analyses.

Other imaging modalities have generally not gained as much popularity. While some report excellent sensitivity and specificity of ultrasound in the diagnosis of appendicitis, the estimates vary significantly. Ultrasound is generally reserved for children and pregnant women for whom the risk of CT scan outweighs the benefit. Magnetic resonance imaging (MRI) is an infrequently used imaging test for suspected appendicitis but does have a role in pregnant women.

Management

In recent years, the question of whether all cases of non-perforated appendicitis requires an emergent trip to the operating room has been called into question. Some case series have demonstrated good outcomes with antibiotic treatment and observation in cases of "mild appendicitis" in patients with Alvarado scores between 4 and 8 and with minimal dilation of the appendix on CT scan. However, trials looking at this exact question have not found the same results and have

demonstrated inferiority of antibiotics to appendectomy for uncomplicated (non-perforated) appendicitis. Specifically, the rate of recurrence in those treated with antibiotics alone was much higher than in the group treated with surgery alone. At the time of this publication, the consensus remains that *non-perforated acute appendicitis warrants a prompt trip to the operating room for appendectomy*.

Advances over the past 20 years in minimally invasive surgery techniques raise the question of whether the appendix is best removed laparoscopically or through the more traditional open approach. This topic has been well studied. In earlier trials, the consensus was that laparoscopy was associated with decreased incidence of wound infection and improved cosmetic result with shorter hospital stays but was associated with increased rates of intra-abdominal abscess postoperatively. In more recent trials comparing laparoscopy with open approaches, laparoscopy was associated with less morbidity, fewer readmissions, and shorter hospitalizations irrespective of severity of disease, including patients with perforated appendicitis at the time of surgery. While not completely undisputed in the literature, multiple retrospective studies have suggested that as surgeons have become more adept at the practice of laparoscopy, in general surgery, the outcomes have improved, and previous critiques of increased abdominal infections, increased time in the operating room, and increased costs are no longer valid.

Management of Complicated Appendicitis

When perforation, abscess, or phlegmon is discovered on preoperative imaging studies, the course of management is often nonoperative, although this too is under constant review in the literature. The mainstay of treatment in these cases is antibiotics, with coverage of enteric gram-negative and anaerobic organisms, irrespective of whether immediate operative intervention is planned. Historically, an identified abscess should undergo percutaneous drainage by interventional radiology. A phlegmon without abscess should be treated with antibiotics. Both should undergo bowel rest until pain resolves. If pain does not resolve, diagnostic laparoscopy allows for additional information gathering and improved diagnostic capability and is indicated in some cases of complicated appendicitis. In recent studies, investigators have begun to consider laparoscopic appendectomy in the acute setting for patients with appendicitis and an associated abscess. Much like the earlier literature looking at uncomplicated appendicitis, laparoscopy is better than open techniques in terms of surgical site infections and length of stay in the hospital but is associated with higher intra-abdominal infections. For patients who do not go to the operating room in the acute setting, interval appendectomy is generally considered 6 weeks after initial presentation, following a period of antibiotics and bowel rest in the acute period.

Special Cases

Pregnant women with appendicitis require special attention. Despite much discussion about the safety of anesthesia for the fetus, the overall rates of fetal loss are fairly low if appendicitis is managed early with surgery before perforation. Overall fetal loss in women with uncomplicated appendicitis is estimated at 2 %, but jumps to 11 % in women with complicated disease. The imaging options to confirm appendicitis in pregnancy include ultrasound, which can be inconclusive, and MRI. While operative intervention in the first trimester has been controversial in the past, the practice around operative intervention in pregnancy has changed and *most favor prompt operative intervention due to the lower rates of fetal loss if appendicitis is managed prior to perforation*.

Conclusion

Appendicitis is as old of a diagnosis as the field of surgery. However, the diagnosis and management of the disease continues to evolve and change both as technology and the ongoing academic investigation of better practices evolves.

43
Diverticulitis

George Singer and Theodore J. Saclarides

Background

Diverticular disease of the colon is an acquired condition affecting individuals with a fiber-deficient diet. Small, hard stools require higher colonic pressures to be propelled caudally. These higher pressures and abnormal motility lead to thickening of the circular muscular layer, shortening of the taeniae, and luminal narrowing. Herniation of mucosa and submucosa then occurs at points of weakness where the vasa recta penetrate the bowel between the taeniae (where there is only a circular muscle layer). Since the sigmoid colon has the smallest diameter of the colon and the highest pressures, diverticula tend to develop here. Erosions of the diverticular wall from increased pressure lead to micro or macro perforations. Colonic bacteria then infect the pericolic fat or mesentery and may progress to localized abscess, diffuse peritonitis, or fistula formation.

Differential Diagnosis

Perforated diverticula produce a range of clinical manifestation from asymptomatic inflammation to generalized peritonitis (Fig. 43.1). Symptoms can include abdominal pain (commonly in the left lower quadrant), fever, chills, and a change in appetite. Other conditions that may initially produce similar symptoms include cancer, appendicitis, gastroenteritis, ischemic colitis, inflammatory bowel disease (IBD), irritable bowel syndrome (IBS), and in females, tubo-ovarian abscess (TOA).

Initial Evaluation

Initial evaluation should begin with a thorough history with attention to the length and progression of symptoms, previous hospitalizations or similar symptoms, cancer-related symptoms such as weight loss and blood in stool, and previous colonoscopy. Physical exam should include assessment of vital signs, palpation and percussion of the abdomen, a rectal exam, a vascular exam, and a pelvic exam in women. A complete blood count should be obtained to evaluate for leukocytosis and anemia. An upright chest X-ray can evaluate for pneumoperitoneum. Antibiotics targeted against gram-negative and anaerobic bacteria should be started. Most physicians use ciprofloxacin or a third-generation cephalosporin and metronidazole; the route of administration (intravenous or oral) is dictated by the patient's clinical condition. A decision must be made whether to continue care on an outpatient basis. Computed tomography (CT) scans of the abdomen and pelvis are frequently obtained early during the illness, especially in the presence of fever or leukocytosis or if the patient requires hospitalization.

Mild Diverticulitis

Patients who are afebrile and without leukocytosis, abdominal tenderness, or pneumoperitoneum may be best treated in the outpatient setting. Treatment consists of oral antibiotics and a clear liquid diet. After resolution of symptoms, patients can be given a low-fiber diet for several weeks (controversial). An outpatient follow-up colonoscopy should be performed 4–6 weeks later to rule out cancer and other causes of the symptoms. The patient should be advised to increase the fiber content of their diet. Approximately 70 % of patients will never have another occurrence.

Peritonitis

Patients with diffuse peritonitis, sepsis, or free pneumoperitoneum are usually gravely ill and require aggressive resuscitation and treatment. Invasive procedures such as flexible sigmoidoscopy, colonoscopy, contrast studies, and CT scans are contraindicated as they only delay laparotomy. Preoperative resuscitation should occur in the emergency room and begin with intravenous (IV) antibiotics and IV fluid. Resuscitation

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_43, © Springer New York 2015

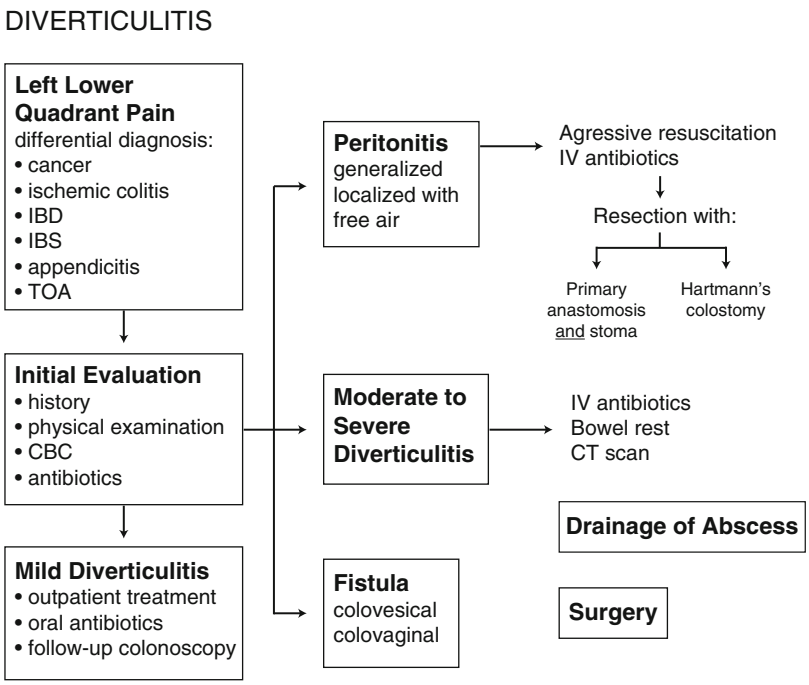

FIG. 43.1 Treatment algorithm for diverticulitis. *IBD* inflammatory bowel disease, *IBS* irritable bowel syndrome, *TOA* tubo-ovarian abscess, *CBC* complete blood count, *IV* intravenous, *CT* computed tomography

should be guided by improvement in heart rate, blood pressure, and urine output. Laparotomy should be performed once the patient is resuscitated. Rupture of a pericolonic or a pelvic abscess can produce purulent peritonitis; free rupture of the colon can cause feculent peritonitis. Management of these patients is determined in the operating room. Options include resection of the diseased portion of the colon and primary anastomosis with diverting ostomy or resection with an end colostomy and mucous fistula or closed rectal stump (Hartmann's operation). The entire abdomen should be irrigated. Colostomy formation and drainage of the abdomen without resection of the diseased portion of the colon is not considered standard management and should be reserved for unstable patients who cannot tolerate a longer operation. However, there is recent literature advocating laparoscopy and peritoneal lavage, proceeding with laparotomy only if feculent peritonitis is found. This approach should be considered investigational. Contrast studies and colonoscopy should be performed in the future, prior to an elective takedown of an ostomy in the future.

Moderate to Severe Diverticulitis

The majority of patients who present to their physician or emergency room do not require emergency laparotomy. Though they may have fever, localized tenderness, or leukocytosis, they can be managed as an inpatient with IV antibiotics, bowel rest, and IV fluid hydration. Nasogastric tubes can be reserved for patients with vomiting from an ileus or obstruction. Failure to improve over 24–48 h should prompt a CT scan to evaluate for abscess formation. CT findings suggestive of diverticulitis include edema of the pericolic fat or mesentery, thickening of the colon wall, and diverticula. Contained extraluminal air does not necessitate a laparotomy; these patients should be managed by their clinical exam. Radiographic-guided percutaneous drainage should be considered for all abscesses greater than 3–4 cm. Inflammatory masses or phlegmons may resolve with antibiotics and time; if not, they may either coalesce to form a drainable abscess in the near future or persist requiring surgery if a prolonged course of antibiotics is unsuccessful.

Fistula

The inflammatory process that accompanies diverticulitis may erode into adjacent organs and structures such as the small bowel, bladder, vagina, and the abdominal wall. Once this occurs, the abdominal symptoms subside and bladder/vaginal symptoms predominate. A colovesical fistula is manifested by pneumaturia, fecaluria, and/or recurrent urinary tract infections. A urinalysis will show fecal material, white blood cells, and bacteria; urine culture will grow enteric bacteria, most commonly *E. coli*. Cystoscopy may be a useful diagnostic modality revealing bullous edema in the dome

of the bladder. A colovaginal fistula can cause fecal material and gas to pass into the vagina, most commonly in women who have undergone prior hysterectomy. Once the inflammatory process has resolved, fistulas are treated with resection of the diseased portion of the colon and primary closure of the adjacent structure, ideally with omentum interposition. A one-stage operation with primary anastomosis is possible for the majority of patients.

Percutaneous Abscess Drainage

Radiographic-guided percutaneous drainage should be considered for all abscesses greater than 3–4 cm that can be accessed safely. Smaller abscesses are likely to resolve with antibiotics and time. The advantage of percutaneous drainage is resolution of the inflammatory process so that the patient may undergo a one-stage resection with primary anastomosis without diverting ostomy. Fistulograms obtained prior to drain removal may demonstrate a fistula to the colon. If a fistula is present, the drainage catheter should be continued and the abscess imaged again when outputs decrease. Alternatively, the catheter is removed at the time of surgery after the patient is stable and pain-free and the inflammatory process has been given time to subside.

Timing of Surgery

Uncomplicated diverticulitis (i.e., no evidence of abscess, fistula, or perforation) is treated nonoperatively as outlined above. Indications for surgery include failure of nonoperative management within a reasonable period of time (usually 5–7 days), worsening clinical signs despite IV antibiotics, bowel rest, and IV fluid resuscitation, recurrent episodes of diverticulitis, and inability to rule out cancer. Surgery for the latter two indications is performed once the inflammatory process has been given time to subside. If cancer is ruled out, the surgeon and the patient must have a discussion regarding the risks and benefits of elective partial colectomy. It is probably not necessary to operate sooner simply because the patient is young, as was formerly thought. Surgery should be considered after two hospitalizations regardless of age or after one hospitalization for a complicated attack (contained perforation, abscess).

The proximal and distal lines of resection are chosen so that the likelihood of recurrent diverticulitis is minimized. This requires the removal of the high pressure areas of the colon, commonly the sigmoid colon and hypertrophied descending colon. The proximal line of resection should be proximal to hypertrophied bowel wall as determined by palpation and visual inspection. The distal line of resection should be the rectum, defined as that area where the taeniae expand to surround the entire rectal wall as the longitudinal muscle layer. Not all areas of the colon with diverticula need to be resected; in fact, to do so would jeopardize the functional result of the operation. A diet high in fiber should reduce the chance of developing diverticulitis again. If the inflammatory process has been severe, the anatomy may be distorted, and preoperative ureteral catheter placement should be considered. While ureteral catheters do not decrease the risk of ureteral injury, they do facilitate identification of intraoperative injuries that can be repaired. If a colostomy is even a remote possibility, stoma sites should be marked preoperatively with consideration of the patient's body habitus, skin creases, waistline, and bony prominences.

44
Large Bowel Obstruction

Kyle G. Cologne

Introduction

In the United States, the most frequent causes of large bowel obstruction (LBO), in decreasing order, include colorectal cancer, diverticulitis (or other inflammatory disorders such as inflammatory bowel disease (IBD)), sigmoid or cecal volvulus, fecal impaction, and then a number of other uncommon causes such as foreign bodies, sliding inguinal hernias, and endometriosis. In children, Hirschsprung's disease, imperforate anus, and meconium ileus are common causes. Finally, colonic pseudo-obstruction should be included in the differential as it has a similar radiologic appearance, namely, proximal colonic distention and a paucity of distal gas. The key to differentiating amongst these is a thorough history and physical and a stepwise approach to ordering additional diagnostic tests, which include both imaging and endoscopic evaluations.

History and Physical Exam

Typical symptoms may include abdominal distention, cramps, and obstipation but may vary widely depending on the level of obstruction and the presence of a competent ileocecal valve (which prevents dilation of the small bowel and creates, in essence, a closed-loop obstruction). Associated symptoms may also provide clues to the underlying etiology (Fig. 44.1).

Patients with *colorectal cancer* represent the most common cause of acute large bowel obstruction encountered in practice today. These patients may give a history of a variety of nonspecific complaints including: weight loss, unexplained anemia, altered stool consistency, bright red blood per rectum, or a family history of colorectal or other cancers (i.e., endometrial, gastric, ovarian, or breast cancer). A strong family history may represent a genetic defect that needs further investigation. Obstruction or perforation signify a more advanced stage or portend a worse prognosis.

Diverticular disease represents the second most common cause of acute large bowel obstruction. A diverticular stricture may develop after a single bout of diverticulitis or patients may give a history of having several previous episodes of pain. Differentiation between this and cancer is often difficult on history alone, and any emergent operation, if needed, should proceed with this in mind. Operative findings may be similar in that both cancer and diverticulitis may cause a mass that is adherent to surrounding structures.

Volvulus represents the third most common cause and includes primarily sigmoid volvulus, but also cecal volvulus, and less commonly other areas. This is much more common in some areas of the world, the so-called volvulus belt, which includes parts of Africa, the Middle East, and Brazil. In these regions, fiber intake is much higher and an elongated sigmoid colon with a short mesentery is common. In these countries, the average age is much younger (fourth or fifth decade) compared to the United States, where a typical patient with volvulus is in the sixth or seventh decade and may reside either in a nursing home or psychiatric institution.

Colonic pseudo-obstruction, or Ogilvie's syndrome, is a nonobstructive dilation of the colon due to underlying dysfunction. It usually occurs in patients who are hospitalized for other reasons such as burns, trauma, or orthopedic, cardiac, or vascular conditions. Patients are frequently taking narcotic pain medications or have electrolyte imbalances. This diagnosis *requires exclusion of an obstruction by contrast enema or endoscopy*, as plain abdominal imaging looks similar to some of the conditions described previously.

Resuscitation

Unlike small bowel obstruction (SBO), large bowel obstruction will often require surgical intervention. Regardless of cause, the first step is always fluid resuscitation, as patients are often intravascularly depleted, especially if they have had a delayed presentation or have been vomiting. A nasogastric tube may be inserted to decompress the stomach and keep swallowed air from worsening the colonic distention. The tube, however, will not decrease the amount of abdominal distention or decrease the amount of colonic gas.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_44,

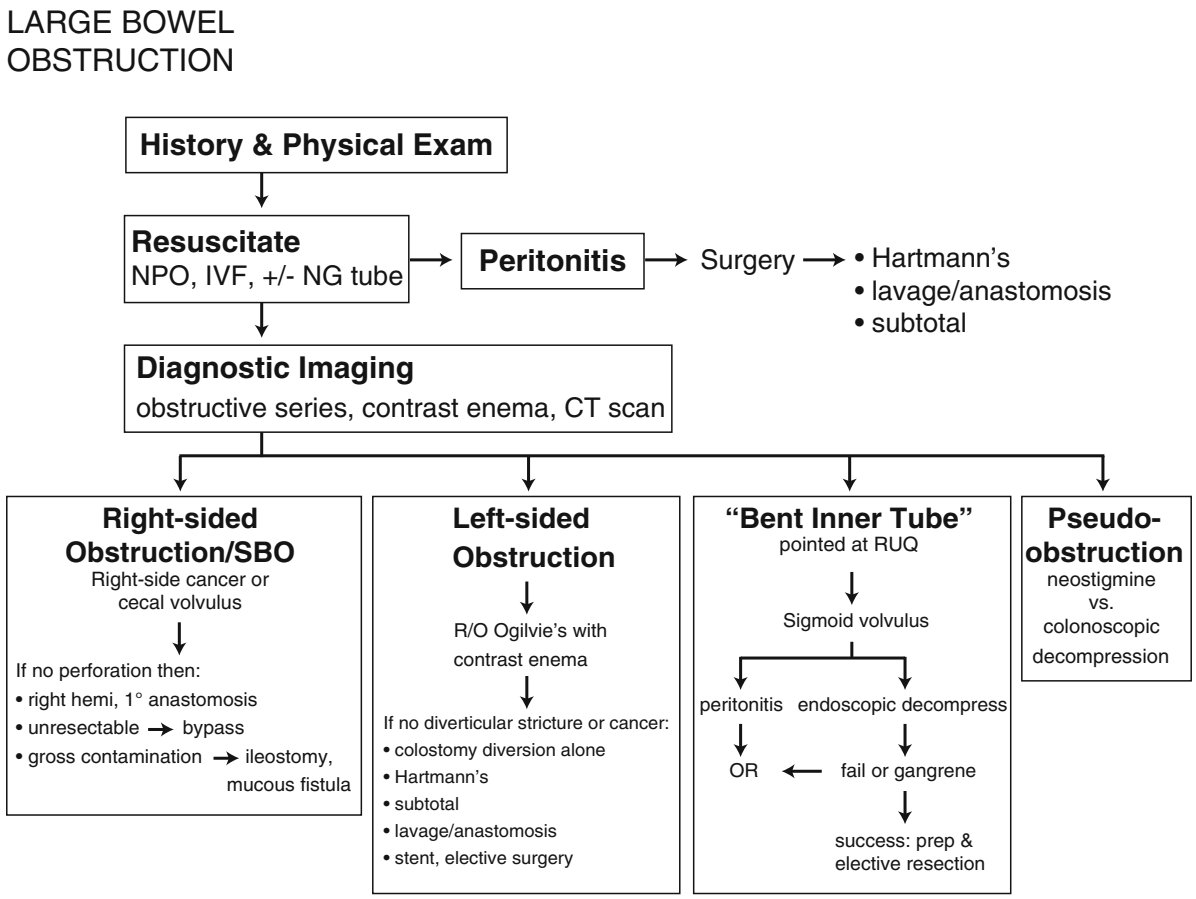

FIG. 44.1 Treatment algorithm for large bowel obstruction. *NPO* nothing per os, *IVF* intravenous fluid, *NG* nasogastric, *CT* computed tomography, *RUQ* right upper quadrant, *SBO* small bowel obstruction

Peritonitis

If signs of peritonitis are present, immediate surgery is indicated. Time spent pursuing radiologic or endoscopic tests only delays laparotomy. The patient should be informed that a colostomy will likely be necessary.

If peritonitis is not present, a stepwise ordering of tests can facilitate a more accurate diagnosis. This also allows time to address any comorbid illnesses, which are often present in colonic obstruction since many of the underlying causes have a tendency to affect older individuals.

Diagnostic Imaging

The first test ordered should be an obstructive series. This may allow determination of the level of obstruction and may make further diagnostic testing unnecessary (e.g., shows free air). If the ileocecal valve is incompetent (seen in 40–50 % of patients), small bowel dilation may make exclusion of a small bowel obstruction difficult. Abrupt cutoff of the colonic gas pattern in the left abdomen with proximal dilation is suggestive of a distal obstructing cancer, diverticular stricture, or pseudo-obstruction. Sigmoid volvulus has a characteristic "bent inner tube" appearance: an air-filled loop that rises out of the pelvis and *points toward the liver*. A cecal volvulus has a "coffee-bean sign" that appears similar but *points toward the spleen*.

According to the law of Laplace, mural tension (pressure) is highest at the point in a cylinder where luminal diameter is greatest, i.e., the cecum. Cecal dilation in excess of 10–12 cm suggests the possibility of impending cecal rupture, and immediate surgical intervention should be taken to avoid compromising survival, as perforation with peritonitis translates into significantly worse outcomes. This is especially true in the setting of a competent ileocecal valve, as there is no backward release of pressure into the small bowel and a closed-loop obstruction may exist. Transverse colon diameter greater than 8 cm should similarly raise concern. If none

of these findings are seen on obstructive series, additional imaging may be obtained. The caveat with these recommendations regards the speed with which this occurs. Dilatation that occurs slowly over a long period—e.g., chronic pseudo-obstruction in Parkinson's—does not carry the same concern for perforation as an acute obstruction.

Since the exact site of the obstruction has implications on the choice for operation, a contrast enema or computed tomography (CT) scan with water-soluble rectal contrast may be obtained. This defines the location and degree of obstruction. CT scan has the additional advantage of showing extraluminal disease, such as metastatic implants in the liver or severe omental involvement (seen as caking or carcinomatosis).

While most acute presentations of large bowel obstruction will be complete, passage of contrast through to a more proximal location suggests a partial obstruction and may also give clues to the nature of the underlying condition. If an area of stricture is seen, the mucosal pattern can suggest etiology. Diverticular stricture typically is a tapered narrowing with preserved mucosal pattern within the stricture, whereas an obstructing cancer has "shoulders" at the point of narrowing and irregular contours within the stricture (the so-called apple-core appearance). A "bird's beak deformity" is seen if a volvulus is present. In colonic pseudo-obstruction, contrast should pass freely to the area of dilation without any stricture, narrowing, or mass effect. It should be performed under fluoroscopic guidance to prevent overdistention of the colon and minimize the risk of perforation.

Right-Sided Obstruction Management

Obstruction of the right colon may present with small bowel distention, or limited colonic distention with a closed-loop obstruction, and usually represents a cancer. A right hemicolectomy and primary anastomosis can be safely performed in most cases unless perforation with a significant amount of contamination has occurred. In this instance, resection with end ileostomy and mucous fistula is preferred. If a cancer is bulky and unresectable, intestinal bypass may be considered as an alternative.

Left-Sided Obstruction Management

Obstruction of the left colon presents a more challenging management decision, since a large column of feces exists above the point of obstruction. It is dealt with by a variety of surgical procedures that include resection and reanastomosis with or without a stoma, resection and colostomy (Hartmann's procedure), and colostomy without resection. The choice depends on the patient's overall physiologic condition, comorbid conditions, and ability to tolerate major surgery. If only a partial obstruction exists and slow bowel preparation followed by elective resection is possible, the patient may undergo a one-stage procedure. Alternatively, an on-table lavage can be performed to eliminate impacted feces from the colon prior to performing an anastomosis. The most commonly performed procedure is the Hartmann's resection, although 30–40 % of these stomas never get reversed. Finally, in the frail, elderly, or unstable patient, a colostomy alone relieves the obstruction while spending the least amount of time in the operating room (provided ischemic bowel is not the source of instability). A resection can always be performed at a later date. In the realm of colon surgery, one of the most morbid complications is an anastomotic leak, and this should be considered in deciding what procedure to perform.

Colonic stenting is an additional option that can be used either as a bridge to elective (rather than emergent) surgery or as a definitive palliative procedure. Stenting can be successfully performed in 80–90 % of cases in experienced hands. The most common complications following stent placement are migration and re-obstruction.

If sigmoid volvulus is present, as suggested by the "bent inner tube" sign on plain X-ray or a "bird's beak deformity" on contrast study, the patient should be assessed for signs of colonic ischemia. These include fever, tachycardia, an elevated white blood cell count, acidosis, or hypotension. These signs would suggest that the patient should be taken for laparotomy. If not, an attempt should be made at endoscopic decompression, with a rigid or flexible endoscope. If successful and if no compromised mucosa suggestive of ischemic bowel is seen, a colonic tube should be left in place to allow bowel prep and a definitive procedure during the same hospital admission. Endoscopic decompression alone results in a recurrence rate of approximately 40–70 %. Sigmoid resection is the most successful option for reducing future recurrences, although in poor surgical candidates, a sigmoidopexy or watch-and-wait strategy may be employed.

Pseudo-obstruction

If the diagnosis of colonic pseudo-obstruction is confirmed, colonic stimulation with the use of neostigmine or colonoscopic decompression is indicated.

Conclusion

In summary, large bowel obstruction has a number of common causes including cancer, inflammatory stricture, and volvulus. The history and plain abdominal imaging can lead to a diagnosis in a significant number of cases. The use of CT or contrast imaging and endoscopic examination allows both diagnosis and treatment of underlying causes. Surgical options depend on the level of obstruction and should be chosen based on the disease process and patient's level of health.

45
Colon Cancer

Abid Khan and Marc I. Brand

Introduction

The implementation of a screening process combined with advances in surgical techniques and chemotherapy have contributed to a nearly one-third decrease in the incidence of and the mortality from colorectal cancer over the past two decades. However, colorectal cancer remains the third most common and third most fatal cancer in the United States. The American Cancer Society estimates that more than 140,000 people were diagnosed with colorectal cancer in 2011 and that nearly 50,000 people died from the disease.

Screening

The decrease in incidence and mortality is primarily a result of an effective screening process for colon cancer (Fig. 45.1). The basis for colon cancer screening is interrupting the *adenoma-carcinoma sequence*. This theory postulates that normal colonic mucosa first transforms into an adenoma and then into cancer over the course of 10 years. The slow transition to carcinoma allows for a manageable screening protocol that has the potential to actually prevent cancer. The National Polyp Study found that individuals undergoing polypectomy had an almost 90 % reduction in the development of colon cancer compared with the average population.

The first step in screening is to assess an individual's colon cancer risk. Risk factors for the development of colon cancer include a personal history of adenomas or colon cancer, inflammatory bowel disease, breast or genital tract cancers, and previous radiation. Several lifestyle factors have been implicated in increasing colon cancer risk, including lower levels of physical activity, increased body mass index, smoking, and a high-fat high-calorie diet. Inherited syndromes account for approximately 5 % of colon cancers, with the most common being hereditary nonpolyposis colorectal cancer (HNPCC) followed by familial adenomatous polyposis (FAP).

An average-risk individual is asymptomatic and has no history of adenomas or colon cancer, has no first-degree relatives with colon cancer, and has no history of inflammatory bowel disease. These individuals should be offered colon cancer screening beginning at age 50 (40 for African Americans). Some have suggested that patients with multiple lifestyle-related risk factors should begin screening earlier; however, no agreed upon guidelines exist.

The current (2008) recommendation from the US Preventive Services Task Force (USPSTF) is for individuals 50–75 years old to undergo one of the following three regimens:

1. Annual high-sensitivity fecal occult blood testing (FOBT)
2. Sigmoidoscopy every 5 years with high-sensitivity fecal occult blood testing every 3 years
3. Colonoscopy every 10 years

The USPSTF recommends against routine screening of individuals 75–85 years old, as the impact of screening on mortality declines with age. The patient's risk, functional status, and a variety of other factors must be evaluated when deciding whether to screen these individuals. The USPSTF recommends against any screening in patients older than 85 years old. CT colonography and fecal DNA testing may prove to be adequate for colon cancer screening, but the current USPSTF recommendations state that further evidence in support of these modalities is needed.

Higher risk individuals require more stringent screening regimens. Patients with a single low-grade adenoma less than 1 cm in size should undergo a follow-up colonoscopy in 5 years. Patients with multiple, high-grade, or large (>1 cm) adenomas should undergo repeat colonoscopies at 3 and 6 years. If these repeat colonoscopies do not detect polyps, these patients should return to a 10-year screening schedule. *If there is question as to whether a high-grade polyp was fully cleared, a patient should undergo a repeat colonoscopy in 2–6 months*. Patients with inflammatory bowel disease should undergo colonoscopy every 1–2 years starting 8 years

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_45, © Springer New York 2015

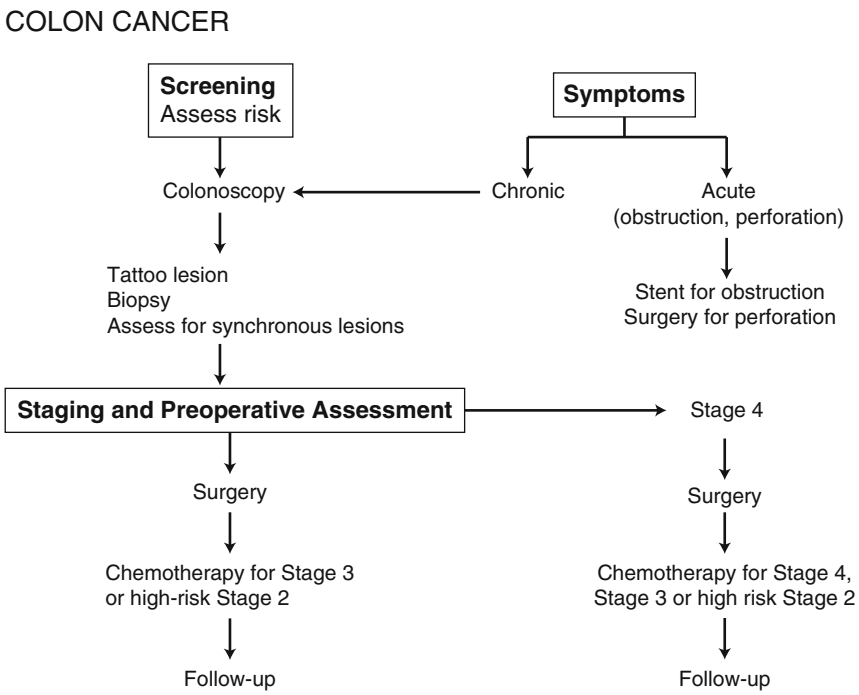

FIG. 45.1 Treatment algorithm for colon cancer

after diagnosis. Those patients with a significant family history (a first-degree relative diagnosed before age 60 or two first-degree relatives at any age) should have their first colonoscopy at 40 years old and repeat every 5 years. Those with one first-degree relative diagnosed after age 60 or two second-degree relatives with colon cancer should have their first colonoscopy at age 40 and then follow the recommendations for average-risk individuals. Patients with suspected FAP should have a yearly flexible sigmoidoscopy beginning at puberty. Those with suspected HPNCC should have a colonoscopy every 1–2 years beginning at age 20 or 10 years younger than the youngest family member's diagnosis.

Preoperative Assessment

Symptoms of colon cancer may be gradual (bleeding, melena, change in bowel habits, pain) or acute (obstruction, perforation).

Due to a 5 % risk of synchronous lesions, a total colonoscopy should be performed whenever possible. In the event that the malignancy prevents endoscopic evaluation of the entire colon, an air-contrast barium enema or CT colonography study should be used to assess the rest of the colon. Alternatively, an obstructing lesion may be opened with a stent, which allows for subsequent colonoscopy. "Tattooing" the colon distal to the tumor allows easier identification of the mass during surgery, especially if laparoscopic surgery is considered. Finally, an assessment of the patient's cardiopulmonary and functional status is necessary to determine if the patient is a candidate for surgery.

Preoperative clinical staging is accomplished through history and physical, endoscopic findings, and biopsy results (Table 45.1). A chest X-ray should be obtained in every patient to look for lung metastasis. A CEA level should be obtained prior to surgery. The role of preoperative CT scan of the abdomen and pelvis remains controversial. Obtaining a preoperative CT scan is routine in some centers; however, evidence supporting this practice is limited. PET scan has proven to be more sensitive than CT in detecting metastasis, but the routine use of this modality is not recommended. Definitive pathologic staging can only be carried out after resection as both nodal and metastatic status are used in staging, as well as in treatment and prognosis.

Surgical Resection

Polypectomy alone is adequate treatment for some T1 cancers: if the margins are negative, the tumor is not poorly differentiated, and there is no lymphovascular invasion. For other colon cancers, the goal in treatment is to perform an R0 resection (no residual disease) with negative margins and an en bloc lymphadenectomy, with harvesting of at least 12 lymph nodes. The major arterial supply to the segment of bowel resected should be ligated as close to its origin as possible. Tumors that invade adjacent organs may require en bloc resection of part or all of the affected organ.

TABLE 45.1. American Joint Committee on Cancer TNM clinical classification of colorectal cancer.

Primary tumor (T)	
TX	Primary tumor cannot be assessed
TO	No evidence of primary tumor
Tis	Carcinoma in situ: intraepithelial or invasion of lamina propria*
Tl	Tumor invades submucosa
T2	Tumor invades muscularis propria
T3	Tumor invades through the muscularis propria into pericolorectal tissues
T4a	Tumor penetrates to the surface of the visceral peritoneum**
T4b	Tumor directly invades or is adherent to other organs or structures**· ***
Regional lymph nodes (N)	
NX	Regional lymph nodes cannot be assessed
NO	No regional lymph node metastasis
N1	Metastasis in 1–3 regional lymph nodes
N1a	Metastasis in 1 regional lymph node
N1b	Metastasis in 2–3 regional lymph nodes
N1c	Tumor deposit(s) in the subserosa, mesentery, or nonperitonealized pericolic or perirectal tissues without regional nodal metastasis
N2	Metastasis in 4 or more regional lymph nodes
N2a	Metastasis in 4–6 regional lymph nodes
N2b	Metastasis in 7 or more regional lymph nodes
Distant metastasis (M)	
MO	No distant metastasis
M1	Distant metastasis
M1a	Metastasis confined to one organ or site (e.g., liver, lung, ovary, nonregional node)
M1b	Metastasis in more than one organ/site or the peritoneum

Anatomic stage/prognostic groups

Stage	T	N	M	Dukes*	MAC*
0	Tis	N0	M0	—	—
I	T1	N0	M0	A	A
	T2	N0	M0	A	B1
IIA	T3	N0	M0	B	B2
IIB	T4a	N0	M0	B	B2
IIC	T4b	N0	M0	B	B3
IIIA	T1-T2	N1/N1c	M0	C	C1
	T1	N2c	M0	C	C1
IIIB	T3-T4a	N1/N1c	M0	C	C2
	T2-T3	N2a	M0	C	C1/C2
	T1-T2	N2b	M0	C	C1
IIIC	T4a	N2a	M0	C	C2
	T3-T4a	N2b	M0	C	C2
	T4b	N1-N2	M0	C	C3
IVA	AnyT	Any N	M1a	—	—
IVB	Any T	Any N	M1b	—	—

Used with permission of the American Joint Committee on Cancer (AJCC), Chicago, Illinois. The original and primary source for this information is the AJCC Cancer Staging Manual, Seventh Edition (2010) published by Springer Science+Business Media. Chapter 14, Colon and Rectum. In: Edge SB, Byrd DR, Compton CC, eds. AJCC Cancer Staging Manual. 7th ed. New York, NY: Springer (page 155)

Laparoscopic colon resections offer comparable oncologic results to open surgery. The patient must be informed of the potential complications, including anastomotic leak and abscess formation. Many surgeons prefer that a patient undergo bowel preparation prior to surgery; however, recent evidence suggests there may be little benefit to this practice.

Perforations require an emergent exploratory laparotomy and usually necessitate an ostomy (either a Hartmann's pouch or a diverting loop ostomy with primary anastomosis). Obstructing lesions proximal to the splenic flexure can often be treated with resection and a primary anastomosis between the ileum and colon. Obstructing lesions distal to the splenic flexure usually require fecal diversion in addition to resection. In some instances, an endoscopic stent can provide palliation in acute obstruction until the patient is optimized for surgery. Stenting allows time to correct comorbid conditions before surgery and to check the remaining colon for synchronous tumors.

Evidence suggests resection of liver or lung metastasis improves survival. Lesions may be resectable if their location permits and RO resection and there is adequate hepatic reserve to maintain life.

Surgery for metastasis may be performed at the time of colon resection or as part of a staged procedure; the latter is preferred if resection requires anything more than a lobectomy. If a patient presents initially with stage 4 disease, systemic therapy (chemotherapy) is the preferred treatment unless the tumor has caused significant bleeding or an obstruction, in which case, resection of the primary tumor is undertaken first.

After Surgery

Early initiation of PO intake, limitations on IV fluids, aggressive pulmonary toilet, and early ambulation are mainstays of post-colectomy care. Pain control should be accomplished with PCA pumps, IV anti-inflammatory medications, and epidural catheters in the initial postoperative period but should be aggressively weaned.

After the immediate postoperative period, attention should turn to cancer management. Any Stage 3 or 4 colon cancer should be treated with adjuvant chemotherapy. It may be considered for high-risk stage 2 cancers (high grade, T4 lesions, obstructing lesions, lymphovascular invasion). The current standard treatment is a combination of 5-fluorouracil (5-FU), leucovorin, and oxaliplatin (FOLFOX).

A patient with colon cancer requires close follow-up. Patients should return for a physical examination every 3–6 months for 3 years and then annually. CEA testing should be performed every 2–3 months for 2 years, then every 3–6 months for 3 years, and then annually. If CEA levels were high prior to surgery and dropped to normal after surgery, a high CEA level at follow-up suggests a recurrence. Colonoscopy should be performed at 1 year after surgery, again at 3 years after surgery, and then every 5 years thereafter. Routine chest X-ray or CT scans are not recommended as part of follow-up.

Conclusion

Over the past 20 years, significant progress in the prevention and treatment of colon cancer has been made, but there is still room for improvement. Novel chemotherapies are being developed and tested with positive early results. Improved genetic testing will allow for more accurate detection of those individuals at highest risk. However, providing access to screening for the entire population will be the best way to limit the burden from this disease.

46
Lower Gastrointestinal Hemorrhage

Benjamin Veenstra and Jonathan A. Myers

Introduction

By definition, a lower gastrointestinal (GI) bleed is one that originates distal to the ligament of Treitz. This leaves a broad anatomical region (small bowel, colon, rectum, and anus) in terms of isolating the source. In children and young adults, the differential diagnosis includes Meckel's diverticulum, intussusception, inflammatory bowel disease (IBD), and juvenile polyps. In adults, common etiologies include diverticulosis, angiodysplasia, neoplasm, IBD, and anorectal disorders. This chapter will focus on hemorrhage in adult patients. Most causes of lower GI hemorrhage originate in the colon (95 %) with diverticular disease being the most common.

Initial Assessment and Resuscitation

Hemorrhage suggests a rapid loss of blood volume and is often associated with hemodynamic instability. Prompt resuscitation of the patient is paramount. The *ABC*s are followed and once *a*irway and *b*reathing are assessed and secured, proper attention can be turned towards *c*irculation. Findings that suggest a significant loss of volume include tachycardia, hypotension, cool and clammy extremities, and altered mental status. Intravenous (IV) access must be obtained quickly, typically with two large-bore IVs (16 G) at the antecubital fossa (Fig. 46.1). A Foley catheter is inserted. A fluid challenge of 2 L crystalloid is administered and baseline labs (including type and cross, CBC, coagulation profile, and CMP) are sent. The hemoglobin is unreliable in an active bleed and a "normal" result should be viewed with caution. If the vital signs respond to the initial crystalloid bolus, this fluid can be continued; otherwise, blood is used for further resuscitation. A period of 12–24 h is often needed for equilibration of the hemoglobin. The intensive care unit (ICU) is the most appropriate setting for continued resuscitation and workup of an acute lower GI hemorrhage.

A focused history and physical is essential and is performed simultaneously with the resuscitation. Relevant history includes prior bleeding episodes, the use of nonsteroidal anti-inflammatory drugs (NSAIDs), ethanol (ETOH) abuse, liver or renal disease, cancer, and coagulopathies. Further characterization of the bleeding episode(s) is also essential. Duration, frequency, and degree of blood loss are important patterns to note. Additionally, associated symptoms including change in bowel movement habits, nausea/vomiting, or hematemesis are relevant.

Nasogastric (NG) lavage and rectal exam with proctoscopy are performed early. The purpose of NG lavage is to distinguish an upper versus lower source of GI hemorrhage. Blood with lavage is suggestive of an upper GI source, while return of gastric contents with bile indicates a lower source. Once a lower GI source is established, a thorough exam of the anus and rectum is necessary. The goal of this exam is to identify any source of distal bleeding whether it is related to a fissure, low-lying rectal mass, or hemorrhoidal disease. This is best completed with digital rectal exam (DRE) and rigid proctoscopy.

Bleeding Pattern and Severity

1. *Occult* blood loss may cause fatigue, anemia, and guaiac-positive stools. Often, no gross blood is noted on exam. Workup is commonly done in an outpatient setting. The differential diagnosis includes colorectal cancer.
2. *Intermittent* blood loss presents as self-limiting episodes of bleeding per rectum. The length and number of episodes vary. Patients typically remain hemodynamically stable and workup is completed either in an inpatient or outpatient setting. The differential diagnosis is vast, ranging from anorectal sources to angiodysplasia to diverticular disease.
3. *Massive* blood loss is often associated with hemodynamic instability. This workup is usually completed in an

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_46, © Springer New York 2015

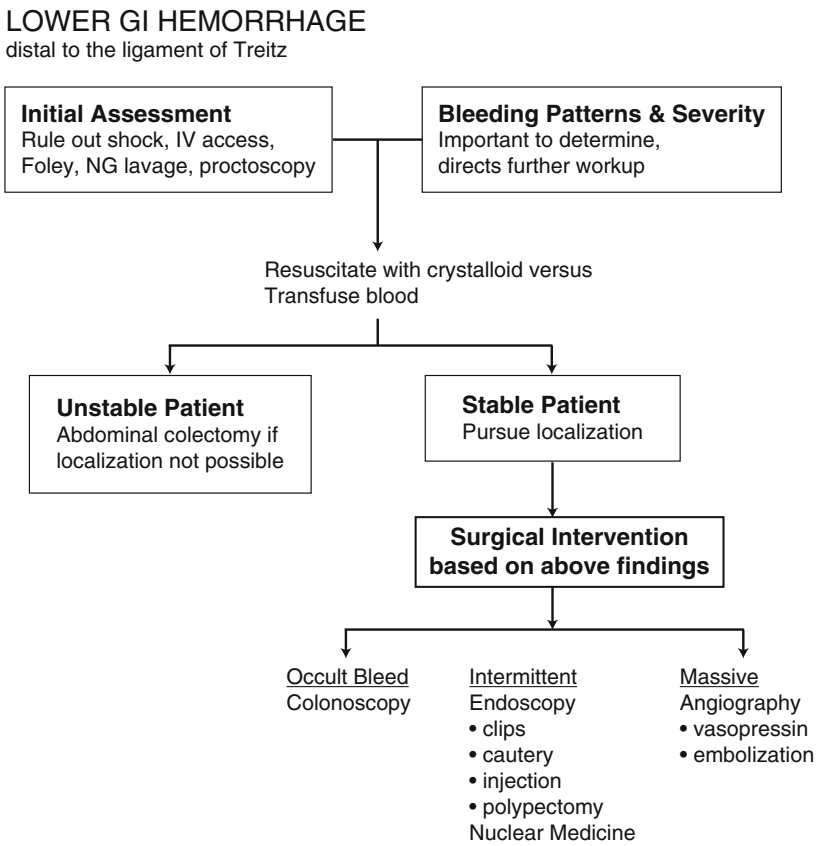

FIG. 46.1 Algorithm for treatment of lower gastrointestinal hemorrhage. *IV* intravenous, *NG* nasogastric

inpatient setting (generally in the ICU) and attempts at localization are done in an expedient manner.

4. *Obscure* blood loss is defined as persistent bleeding after negative attempts at localization, most commonly esophagogastroduodenoscopy (EGD) and colonoscopy. It can be further divided into obscure occult and obscure overt. In obscure occult, blood loss manifests as anemia and guaiac-positive stools. Obscure overt is more consistent with an intermittent bleeding pattern.

Treatment of the Unstable Patient

Localization of a lower GI hemorrhage source is essential prior to operation. However, at times, it is necessary to operate emergently without localization, depending on the clinical setting and the severity of the bleeding. The operation of choice is a total abdominal colectomy leaving the rectum. Either an ileostomy or an ileorectal anastomosis is created. Since precise localization of the bleeding site was not performed, a segmental colectomy is not advised. The small bowel is inspected at the time of surgery.

Treatment of the Stable Patient

In the stable patient, localization is essential in performing the correct operation. Many of the localization modalities (colonoscopy and angiography in particular) can be therapeutic as well as diagnostic. If a bleeding source is identified and nonoperative management is not possible or is unsuccessful, surgery is indicated. Localization allows for a segmental resection of the colon, thus avoiding the morbidity of a blind total abdominal colectomy. Below is a summary of each of the bleeding patterns and the particular diagnostic/therapeutic tests that can be used to aid in localization of a lower GI hemorrhage.

1. Occult Bleed – *Colonoscopy* is the diagnostic test of choice. A thorough examination of the colon and anorectal region is performed. Many polyps can be removed; larger masses can be biopsied to exclude cancer.
2. Intermittent Bleed – Again, *colonoscopy* is the diagnostic test of choice. This is performed in between episodes of bleeding if possible or at a point when the bleeding has slowed. An adequate prep is needed in order to fully visualize the entire colon. In the proper setting, colonoscopy is successful in localizing a bleeding source in up to 95 % of patients. If an active bleed is noted, attempts can be made endoscopically to control it. Modalities that are used include clips, injection of vasoconstricting agents, and thermal ablation. If an active bleed is identified and cannot be controlled endoscopically or the patient rebleeds post-endoscopic therapy, surgery is indicated.

   If colonoscopy fails to identify a source but intermittent bleeding persists, a *99Tc-tagged RBC scan* is a logical subsequent exam. The purpose of this test is twofold. First, this exam allows for potential localization.

A tagged RBC scan can detect bleeding as slow as 0.1 mL/min with a reported sensitivity of more than 90 %. However, the accuracy of localization with this method has been disappointing, with reports of 40–60 %. The reason behind this being poor is inexact spatial concordance with the resultant imaging. For example, if a blush is noted in the right lower quadrant, this does not ensure the lesion lies in the ascending colon. The blush could be originating from a Meckel's diverticulum in the small bowel or diverticula in redundant sigmoid colon. The second and more commonly used indication for tagged RBC scan is determining the utility of an angiogram. If the scan is negative or positive after a few hours, an angiogram is unlikely to be sensitive enough to diagnose the location of bleeding.

3. Massive Bleed – The previous two localization modalities are often fraught with difficulty during an ongoing, massive bleed. Visualization during colonoscopy may be impossible in a blood-filled colon. With ongoing blood loss, *selective angiography* is the test of choice. This exam is very specific and can detect hemorrhage at rates of 0.5–1.0 mL/min. Given the propensity for the right colon to be the source of hemorrhage, the superior mesenteric artery is injected first, followed by the inferior mesenteric and celiac arteries. Diverticular bleeds are more likely to show dye extravasation, as they are arterial in nature. Angiodysplasia generally is characterized by the presence of a vascular tuft, as well as early filling and slow emptying of the mesenteric vein. Angiography is also a valid and successful option in terms of nonoperative management. Selective intra-arterial vasopressin infusion (0.2 Units/min) stops bleeding in 80–90 % of patients. However, once vasopressin is discontinued, as many as 50 % rebleed. Another option is embolization of the bleeding vessel with absorbable gelatin strips or coils. A potential complication of this technique is ischemia, given the limited collateral circulation of the colon. However, recent data suggests that this technique can be safely applied.
4. Obscure Bleed – Perhaps the most frustrating and challenging problem related to lower GI hemorrhage is the obscure bleed. Colonoscopy is reattempted to start. If this fails, a tagged RBC scan can assess the utility of angiography. Recent attempts have been made at provocative testing with favorable results. *Provocative testing* involves the administration of fibrinolytics, anticoagulants, or vasodilators during angiography to increase hemorrhage in hopes of localizing a source. If this is negative, attention is turned towards the small bowel. *Push endoscopy* uses a pediatric scope to reach anywhere from 50 to 70 cm past the ligament of Treitz. The success rate of identifying lesions is in the range of 40 %. *Pull endoscopy* utilizes a technique in which a scope with a balloon on the end permits for normal small bowel peristalsis to carry the scope to distal small bowel. This technique is both time-consuming and tiresome. It has largely been replaced by *video capsule endoscopy (VCE)*. With VCE, the patient swallows a small capsule containing a video camera. Images from this capsule are taken as it passes through the entire GI tract. These images are reviewed and, based on time and location, a source of bleeding can be identified. Recent data suggests success rates as high as 90 % with VCE.

Surgery

Ideally, surgery is reserved for those who have been localized through the aforementioned diagnostic modalities. In these cases, a segmental resection is appropriate with a reasonable expectation that rebleeding will not occur. If localization is not obtained, a total abdominal colectomy is the procedure of choice. Although it is thought that the majority of colonic bleeding originates from the right colon, a blind right colectomy results in unacceptable rebleeding rates. It should be additionally noted that a large majority of bleeding stops without intervention or localization. These patients are observed and discharged when stable. However, these patients often experience recurrent bleeding episodes requiring readmission to the hospital. If this occurs, it is important to repeat the algorithm with each readmission.

47
Genetic Predisposition to Colorectal Cancer

Kristin Gross and Marc I. Brand

Introduction

Colorectal cancer is the most common malignancy of the gastrointestinal tract and is the second leading cause of cancer death when both genders are considered. There are several risk factors associated with an increased incidence of colorectal cancer, such as advanced age, high animal fat and low fiber diet, tobacco smoking, pelvic irradiation, and inflammatory bowel disease. However, one must be aware of a special population of patients with increased risk of colorectal cancer due to hereditary syndromes and manage them accordingly.

Recognition of Hereditary Colorectal Cancer Syndrome (HCRCS)

The first step in the management of hereditary colorectal cancer syndrome (HCRCS) is recognizing when the condition is present. Some patients who are asymptomatic are seen for an unrelated condition. Inquiring about a family cancer history and reviewing prior surgical and pathology reports are crucial components in recognizing HCRCS (Fig. 47.1). Many patients with HCRCS will be the first person in a family diagnosed with the syndrome; these patients are called "probands." Other clues may suggest the presence of HCRCS, such as unusual symptoms (abdominal pain, change in bowel habits, blood in stool) or findings (desmoid tumor, skull osteomas, supernumerary teeth) in a young person.

Hereditary nonpolyposis colon cancer (HNPCC) does not have unique clinical manifestations. Its clinical diagnosis is suggested by the "Amsterdam Criteria," also known as the "3-2-1-0 rule": 3 relatives with colorectal cancer, spanning 2 successive generations, with 1 person diagnosed before age 50, and familial adenomatous polyposis (FAP) is excluded. These criteria are very stringent, meant to reduce false positives, but it does create false negatives. To help identify more potential patients, the Bethesda Guidelines were set forth to raise suspicion for the presence of HNPCC, but they are not diagnostic of HNPCC.

Familial Adenomatous Polyposis (FAP)

FAP is a polyposis syndrome related to an autosomal dominant mutation in the APC tumor suppressor gene on chromosome 5q – a gene involved in cell cycle regulation and cell adhesion. Twenty percent of patients with FAP have no known family history of FAP and are described as "spontaneous FAP" cases. Patients with FAP have adenomatous polyps that begin to develop in puberty. Colonoscopy reveals hundreds to thousands of polyps covering the mucosal surface of the colon and rectum. All patients develop cancer by age 40 (100 % lifetime risk). Total colectomy with ileorectal anastomoses or total proctocolectomy with ileoanal pouch reconstruction or end ileostomy is advised. The timing of surgery is based on development of polyp burden and age of the patient. Surgery is usually performed at the patient's earliest convenience (school, graduations, etc.) unless cancer is present, in which case, prompt intervention is advised. Following resection, lifetime proctoscopic surveillance of the residual rectum (if present) should be performed at least annually to look for polyps or tumors. Upper gastrointestinal surveillance is also advised.

Patients with FAP can also develop duodenal adenomas, usually periampullary. Esophagogastroduodenoscopy (EGD) should be performed at least every 5 years to check for duodenal polyps starting at age 25–30 years. Surveillance interval is based on polyp burden at the last EGD. Following colectomy, periampullary duodenal cancer is the most common cause of death with FAP. Desmoid tumors (e.g., mesenteric mass) also occur in patients with FAP; if involvement of the mesentery is significant, resection

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_47, © Springer New York 2015

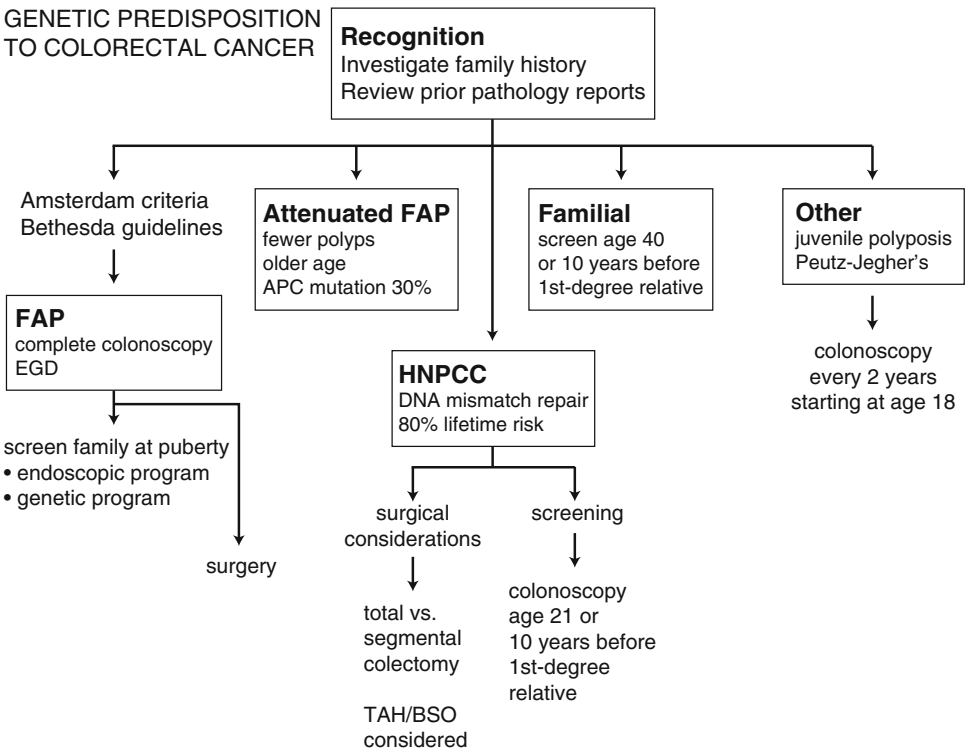

FIG. 47.1 Algorithm for diagnosing and treating genetic predispositions to colorectal cancer. *FAP* familial adenomatous polyposis, *EGD* esophagogastroduodenoscopy, *HNPCC* hereditary nonpolyposis colon cancer, *TAH/BSO* total abdominal hysterectomy and bilateral salpingo-oophorectomy

is not indicated due to the high incidence of short bowel syndrome following resection and high recurrence rate. Because of the high risk associated with surgery, sulindac (COX-2 inhibitor) and tamoxifen may be used initially to induce regression of desmoid tumors.

A diagnosis of FAP requires complete colonoscopy and biopsy to confirm the adenomatous nature (versus hamartomas) of the polyps and to attempt to exclude cancer. Genetic counseling should be offered to the patient's family. Flexible sigmoidoscopy of first-degree relatives is indicated beginning at age 10–15 years to check for polyps. If genetic screening is chosen and APC testing is negative, the relative can be screened at age 50 per average-risk guidelines.

Attenuated Familial Adenomatous Polyposis (AFAP) or MYH-Associated Polyposis (MAP)

Attenuated familial adenomatous polyposis (AFAP) is a variant of FAP and is associated with mutations at the 3′ or 5′ end of the APC gene. Patients present later in life and with fewer polyps, usually 20–100, predominantly in the right colon. Colorectal carcinoma occurs later (average age 55 years) and develops in more than 50 % of patients. Similar to FAP, patients are at risk for duodenal polyposis.

APC gene mutations are present only in about 30 % of patients with attenuated FAP, and when present, they are expressed in an autosomal dominant pattern as in classic FAP. If surgery is indicated due to large polyp burden not manageable with endoscopy, patients are often candidates for a total abdominal colectomy with ileorectal anastomosis because the limited polyposis in the rectum can usually be controlled by colonoscopic snare excision.

In 2002, a new mutation in the MYH gene on chromosome 1p was found to be associated with an increased risk of colonic polyposis and colorectal cancer. MYH is a base excision repair gene. Most patients with MYH-associated polyposis (MAP) exhibit phenotypic overlap with attenuated FAP, with 10–15 adenomas usually found in the fourth decade of life. Unlike APC gene mutations that are expressed in an autosomal dominant pattern, MYH mutations are inherited in an autosomal recessive fashion and the hereditary nature may not be as obvious. Biallelic carriers have an 80 % lifetime risk of colorectal cancer by age 70 years. Patients with MAP should undergo colonoscopy every 1–2 years starting between the ages of 25 and 30 years. Surgical management is similar to AFAP.

Hereditary Nonpolyposis Colon Cancer (HNPCC)

Hereditary nonpolyposis colon cancer (HNPCC) is a syndrome present in 5 % of patients with colorectal cancer. It involves a defect in a DNA mismatch repair gene (hMSH2, hMLH1, hPMS1, hPMS2). This syndrome is inherited in an autosomal dominant pattern. Unlike FAP, these patients do not form thousands of polyps, hence the term "nonpolyposis." These patients have an 80 % lifetime risk of developing colorectal cancer. There is a predilection for right-sided (e.g., cecal) and multiple colon cancers versus sporadic colon cancer where the most common site is the sigmoid colon. Two generalized types of HNPCC have been described, with type 1 assuming the increased colon cancer risk and type 2 with additional cancer risks as well (ovarian, endometrial, bladder, stomach, and pancreas).

At-risk HNPCC patients need surveillance colonoscopy starting at age 21 or 10 years before a first-degree relative acquired cancer, at an interval of every 2 years until age 40, and then every year thereafter. Since patients often have multiple primaries and 50 % get metachronous lesions within 10 years, total colectomy should be performed with the first colon cancer operation. However, genetic confirmation of the HNPCC diagnosis is not always possible before colectomy.

Patients need a surveillance program for extraintestinal malignancies as well. For example, women need endometrial monitoring with ultrasound every 3 years and annual pelvic exams, as well as earlier mammograms than the general population. Women may be offered prophylactic total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH/BSO) after childbearing years if the diagnosis has been confirmed.

Familial Colorectal Cancer

Nonsyndromic colorectal cancer accounts for 10–15 % of patients with colorectal cancer. The lifetime risk of colorectal cancer in the average-risk population is approximately 6 %. This risk increases to 12 % if one first-degree relative is affected and up to 35 % if two first-degree relatives are affected. Age of onset also affects risk, as a diagnosis before age 50 years in a first-degree relative is associated with a higher incidence in family members. Screening colonoscopy for first-degree relatives is recommended every 5 years beginning at age 40 years or 10 years before the age of the youngest diagnosed relative.

Other Hereditary Syndromes Associated with Colorectal Cancer

Juvenile Polyposis Syndrome

Juvenile polyposis syndrome is associated with gastrointestinal hamartomatous polyps and a 10 % lifetime risk of colorectal cancer. The syndrome may be inherited in an autosomal dominant fashion with two associated genes (BMPR1A and SMAD4). Colonic surveillance is indicated every 2 years starting at age 15–18 years. There is no indication for prophylactic colectomy; total colectomy is likely the best option if cancer develops.

Peutz-Jegher's Syndrome

Peutz-Jegher's Syndrome involves gastrointestinal hamartomatous polyposis and dark pigmentation around mucous membranes. These polyps can cause obstruction, with a common first presentation being intussusception. There is still controversy over whether or not a significant increase in the risk of colorectal cancer exists, and prophylactic colectomy is not indicated. However, these patients are at higher risk of extraintestinal malignancies (e.g., pancreas, liver, lungs, breast, ovaries, uterus, testicles). The most common cancer in these patients is breast cancer. Screening for these patients includes EGD and colonoscopy every 2 years starting at age 18 years. Patients also need a screening program for the uterus, ovary, cervix, breast, and testicles.

FAP Variants

Turcot's Syndrome

Turcot's syndrome is an FAP variant associated with the APC gene. Patients develop colon cancer as well as brain tumors.

Gardner's Syndrome

Another FAP variant is Gardner's syndrome, also associated with the APC gene. These patients develop colon cancer, osteomas (bumps on forehead or other bony protuberances, which are benign), and desmoid tumors. As in FAP, these desmoid tumors are benign and not typically amenable to wide local excision due to local extension.

48
Rectal Cancer

Purvi P. Patel and Theodore J. Saclarides

Epidemiology

Last year, approximately 140,000 new cases of colorectal cancer were diagnosed, of which 28 % were in the rectum. The rectum is defined as the area that extends from the rectosigmoid junction (where the taenia coli band together) to the anorectal junction, about 3.5 cm from the anal verge. This area measures approximately 12–16 cm and is divided into the lower rectum (3.5–8 cm), the middle rectum (8–12 cm), and the upper rectum (12–16 cm). From an oncologic and treatment standpoint, tumors in the mid to distal rectum are grouped together and considered distinct from upper rectal lesions. The histological types of rectal cancer include squamous cell carcinoma (SCC), neuroendocrine carcinoma, sarcoma, lymphoma, and adenocarcinoma, the most common subtype and the focus of this chapter.

History and Physical

At presentation, a physician should complete a detailed history and physical examination (Fig. 48.1). History should include a personal and family history of cancer (specifically colorectal cancer), history of polyps, and details regarding change in bowel habits, blood per rectum, or obstructive symptoms. Physical exam must include a digital rectal exam, which can provide details such as tumor size, position, and fixation of the mass. Additional focus should be placed on the abdominal exam for a palpable mass, hepatomegaly, and the presence of inguinal lymphadenopathy. Routine labs including liver function panel and carcinoembryonic antigen (CEA) should be sent. CEA levels can be followed in the postoperative period, with a spike in levels raising concern for recurrence.

Workup

Evaluation should include rigid proctoscopy, colonoscopy, transrectal ultrasound, and additional radiologic imaging to rule out systemic disease. A rigid proctoscope is the best tool to determine tumor location and orientation. This allows one to take reliable and accurate measurements from the anal verge that are essential in establishing the treatment plan. By using a rigid instrument, one can avoid the pitfalls of a flexible scope, which may bend or loop in the rectum giving inaccurate measurements. Colonoscopy should be completed to evaluate the rest of the colon for additional polyps or tumors. An estimated 5–8 % of patients with rectal cancer will have a synchronous cancer and 20–30 % will have a benign polyp elsewhere.

Clinical Staging

Locoregional staging to determine tumor depth and lymph node status is best completed by transrectal ultrasound (TRUS) and/or magnetic resonance imaging (MRI). Ultrasound generates a multilayer representation of the rectal wall marking out the mucosa, submucosa, muscularis propria, and perirectal fat. The stage of the tumor is determined by which layers are disrupted. Features suggestive of lymph node metastasis include spherical shape, hypoechoic nature, and size greater than 1 cm, although some regard visible hypoechoic nodes of any size as suspicious for cancer. The advantages of transrectal ultrasound (TRUS) include its mobility, low cost, good patient tolerance, and no sedation requirements. Its main disadvantage is reliance on operator experience. MRI with endorectal coil is an alternative to TRUS for locoregional staging. MRI provides details regarding mesorectal invasion and surrounding tissue planes.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_48, © Springer New York 2015

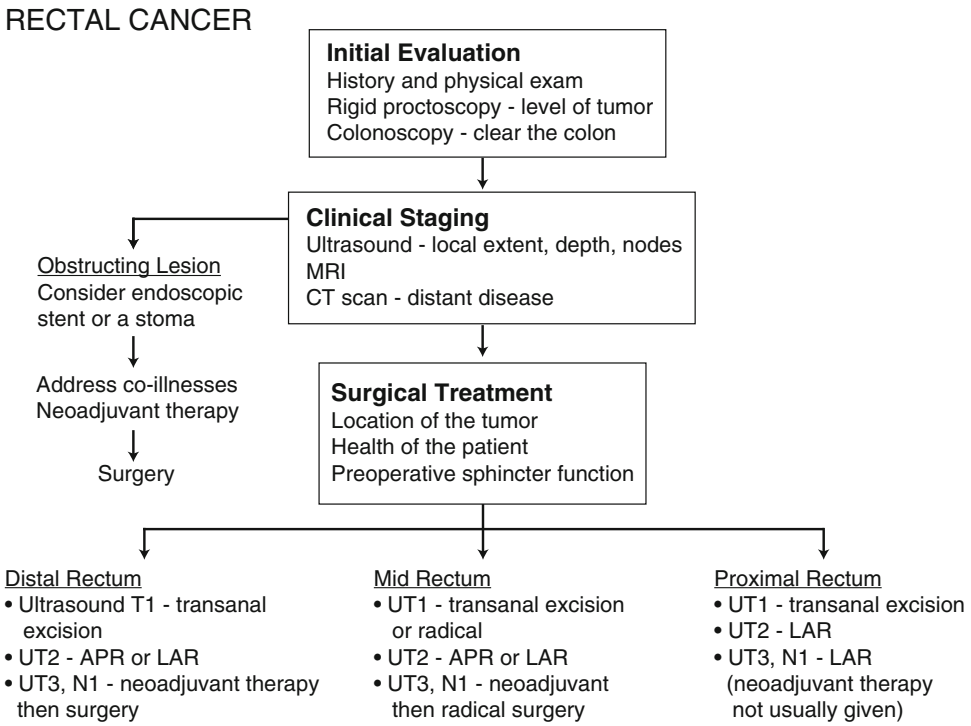

FIG. 48.1 Treatment algorithm for rectal cancer

It is essentially equivalent to TRUS in evaluating tumor depth and nodal disease; however, it is less operator dependent and allows evaluation of near-obstructive lesions.

Distant disease is evaluated by computed tomography (CT) and positron emission tomography (PET). Patients with newly diagnosed rectal cancers usually undergo CT of the abdomen and pelvis. Although this is inferior to both ultrasound and MRI for local staging, it is very good at detecting distant metastatic disease. The most common sites of metastasis are the liver and lungs. CT of the chest may also be included; however, chest X-ray is an acceptable method to evaluate the lungs, followed by CT if any abnormalities are spotted. PET detects fludeoxyglucose, which has increased uptake in metabolically active tissues such as cancer and is usually used as an adjunct to CT.

The information gathered from this evaluation will allow clinical staging of the rectal cancer. The most commonly used pathologic staging system is the TNM system introduced by the American Joint Committee on Cancer and the International Union Against Cancer. "T" quantifies the local depth of tumor invasion, "N" signifies the involvement of lymph nodes, and "M" marks the presence of distant disease. A T1 lesion invades the submucosa while a T2 lesion invades the muscularis propria. Tumors are designated T3 if they invade surrounding perirectal tissues and T4 if there is direct extension into another organ or structure. Regarding nodal disease, the designation N0 means that no nodal metastasis are present, N1 when there are 1–3 regional lymph nodes, and N2 when 4 or greater nodes are seen. The presence of metastatic disease is designated as M1. A patient is initially staged by the radiologic studies and then is reassessed based on the pathologic specimen after surgery.

Surgical Treatment

Neoadjuvant (preoperative) chemotherapy and radiation are recommended for patients with advanced tumors (T3, T4) in the mid and distal rectum, or positive nodal disease seen on ultrasound or MRI. Preoperative chemoradiation has decreased local recurrence rates, downstaged tumors, allowed easier resection of previously bulky tumors, provided better patient tolerance with fewer toxicities, and has increased rates of sphincter preserving surgery. It is recommended that patients who had neoadjuvant chemoradiation receive postoperative chemotherapy. The current chemotherapy recommended is a combination of 5-flurouracil, leucovorin, and oxaliplatin commonly referred to as FOLFOX. This can be given as a slow infusion over several weeks or as a bolus over several days. Neoadjuvant therapy is not administered for cancer in the upper rectum. Postoperative chemotherapy and radiation is reserved for patients with tumor invasion into the perirectal fat or positive lymph nodes who did not receive neoadjuvant therapy. Standard radiation

therapy regimen includes 25 fractions of 1.8 Gy to the whole pelvis with a possible 5.4–9.0 Gy boost to the site of the primary tumor.

Surgical options range from local excision to radical resections. Treatment choice is driven by the clinical stage, tumor location and features, and patient factors. Comorbidities, gender, body habitus, baseline anal tone and function, and personal preferences regarding an ostomy must be considered when creating a surgical plan. Those with multiple medical problems may not be able to tolerate the stress of a long surgery. If a patient already has sphincter dysfunction, attempting sphincter preservation may not be desirable. Since it is essential to obtain a distal margin of at least 2 cm, *tumor location is key in determining if sphincter preservation is feasible*. Tumors of the distal to middle rectum are more likely to require resection of the anal sphincters in order to obtain appropriate margins than those of the upper rectum. Regardless of the procedure, the goals of radical surgical resection include obtaining negative distal and circumferential margins, level-appropriate mesorectal excision, and removal of a minimum of 12 lymph nodes.

Local excision is an appropriate choice for small tumors with favorable histology, limited to the submucosa. The tumors should be well differentiated, lack lymphovascular invasion, and lack perineural invasion. This method entails full-thickness excision with negative margins and primary closure of the defect. Transanal endoscopic microsurgery (TEM) may be employed. TEM procedures use a 4 cm diameter scope with specially designed instruments to allow access to tumors up to 15 cm. The rectal lumen is distended by carbon dioxide insufflation to improve visibility.

Radical surgical approaches involve removing the tumor en bloc with surrounding tissues and lymphatics and allows for intraoperative evaluation of the liver and peritoneum for metastatic disease. A low anterior resection (LAR) involves removal of the rectum with primary anastomosis and is usually possible for cancers in the mid to upper rectum. Functional results following a LAR and low anastomosis may be improved by creation of a fecal reservoir (e.g., colonic J pouch or coloplasty) that may reduce stool frequency, urgency, and tenesmus. Patients undergoing a LAR may require a temporary diverting stoma if the anastomosis is low in the pelvis, if the patient has undergone neoadjuvant chemoradiation, or if a colonic pouch was created. Stoma takedown is completed at a second surgery about 4 months later, after postoperative chemotherapy is completed. An abdominoperineal resection (APR) is reserved for the lowest-lying tumors in which complete excision and negative margins requires removing the rectum, anal canal, and sphincter muscles. This surgery is approached from both the abdomen and perineum and leaves the patient with a permanent stoma. Both LAR and APR involve a level-appropriate mesorectal excision, i.e., removal of lymph node bearing tissue without violating the tissue planes. In the case of an APR, the mesorectal excision is complete and total. A limited total mesorectal excision (TME) dissection to 5 cm below the mesenteric level of the tumor is acceptable for upper rectal cancers. A distal mural margin of 1–2 cm is considered acceptable. Lastly, if the appropriate tissue has been removed, the pathologist must identify a minimum of 12 lymph nodes in the specimen to properly stage the nodal status of the tumor.

Currently, the aforementioned procedures are being completed either with open surgery or laparoscopically. Research has demonstrated equal outcomes in the hands of experienced surgeons in both groups from an oncologic standpoint for colon cancer with ongoing research in its application for treatment of rectal cancer. Laparoscopic surgery has shown a significant reduction in hospital stay, decreased use of narcotics, and earlier return of bowel function. Common complications of LAR and APR include sexual dysfunction specifically retrograde ejaculation and impotence, neurogenic bladder, wound infection, and delayed healing of the perineal wound. These are increased with APR secondary to the extensive pelvic dissection required for adequate resection.

Multidisciplinary teams including surgeons, oncologists, radiologists, radiation oncologists, and pathologists meet to address complex cases (e.g., metastatic or symptomatic disease) and create individualized plans. A single focus of metastatic disease may be considered resectable as part of a staged surgery where the primary tumor is removed first and resection of the metastatic disease is completed several weeks later. These cases may also benefit from chemoradiation in the preoperative or postoperative setting. Patients who present with advanced disease causing an obstruction usually required immediate intervention. Endorectal stents can be placed to relieve the obstruction or a surgically fashioned stoma can be constructed. This will allow time for operative planning, correction of comorbid conditions, and chemoradiation depending on the stage of the disease.

Patients treated for rectal cancer require close postoperative follow-up. In the first 2 years, patients should be seen in the clinic and have a digital rectal exam every 3 months. During these visits, routine labs including liver function panel and carcinoembryonic antigen should be evaluated. Many surgeons also perform a rigid proctoscopy to assess for anastomotic recurrence. Colonoscopy is routinely performed at 1 year after the operation.

49
Anal Cancer

Jacquelyn S. Turner

General Considerations

Cancer of the perianal skin and anal canal are uncommon malignancies, constituting less than 5 % of cancers of the lower gastrointestinal tract. Most cancers in this region arise cephalad to the dentate line of the anal canal by a ratio of 3 to 1. Anal canal cancers are more frequently found in women and anal margin cancers are more frequently found in men. Regardless of location, the patients are usually in their mid-50s when the diagnosis is established [1, 2].

Anatomy of Anal Canal

Cancers in the anal canal are distinctly different from cancers of the anal margin in terms of prognosis and management. Therefore, it is important to understand the anatomy of both locations. The "surgical" anal canal is defined as the terminal part of the large intestine. The anorectal ring (composed of the levator ani muscles) is the proximal part of the anal canal, while the distal part of the anal canal is the anal verge [2]. The mean length of the anal canal is 2–4 cm. Within the anal canal is the transition zone that is about 0–12 mm in length and extends from the dentate line to the tops of the columns of Morgagni. The transition zone has transitional epithelium, which is a cuboidal "urothelium-like" epithelium that is different from the rectal columnar mucosa. The anal margin by definition contains skin appendages, such as hairs. The anal margin extends from the anal verge to about 5–6 cm radial from the anal verge (Fig. 49.1) [1, 2].

Understanding the lymphatic drainage patterns is equally important in the management of anal cancers. Lesions above the dentate line drain to the superior rectal lymphatics and on to the inferior mesenteric and internal iliac lymphatics. Lesions at the dentate line drain to the internal pudendal, internal iliac, and obturator lymph nodes. Lesions below the dentate line drain into inguinal lymph nodes. These lesions may also involve the inferior or superior rectal lymph nodes [1, 2].

High-Risk Conditions

Certain conditions may place an individual at higher risk for squamous cell anal cancer (Fig. 49.2). Immunosuppression from either human immunodeficiency virus (HIV) infection or immunosuppressive medications (transplant patients) has a higher relative risk. In a manner analogous to cervical cancer, infection with the human papilloma virus (HPV), types 16 and 18, raises the risk. The virus can enter through a break in mucosa (i.e., intercourse, friable hemorrhoids, firm stools). Tissue above the dentate line is more susceptible to HPV infections. Other risk factors include cigarette smoking, chronic untreated perianal disease (such as fistulas and Crohn's disease), low CD4 counts, and men who have sex with men. These risk are additive. Currently, routine papanicolaou (pap) smears are advocated by some authors; however, in general, anal pap smears are considered investigational as a method for detection and prevention of anal cancers [1].

Signs and Symptoms

Presenting signs and symptoms include pain, bleeding, itching, fecal incontinence, mucous discharge, and the sensation of a mass. Advanced lesions may impede the passage of stool, erode the vagina, cause tenesmus, invade the sphincter, or metastasize to inguinal lymph nodes. Frequently, an unsuspected cancer will be found within a surgical hemorrhoidectomy specimen [1, 2].

Diagnosis

It is imperative that any unusual, abnormal, or persistent perianal or anal lesion be biopsied to establish a diagnosis. This is especially so for nonhealing lesions and lesions that are refractory to conservative measures. Tissue may be obtained by a simple office-based biopsy under local

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_49, © Springer New York 2015

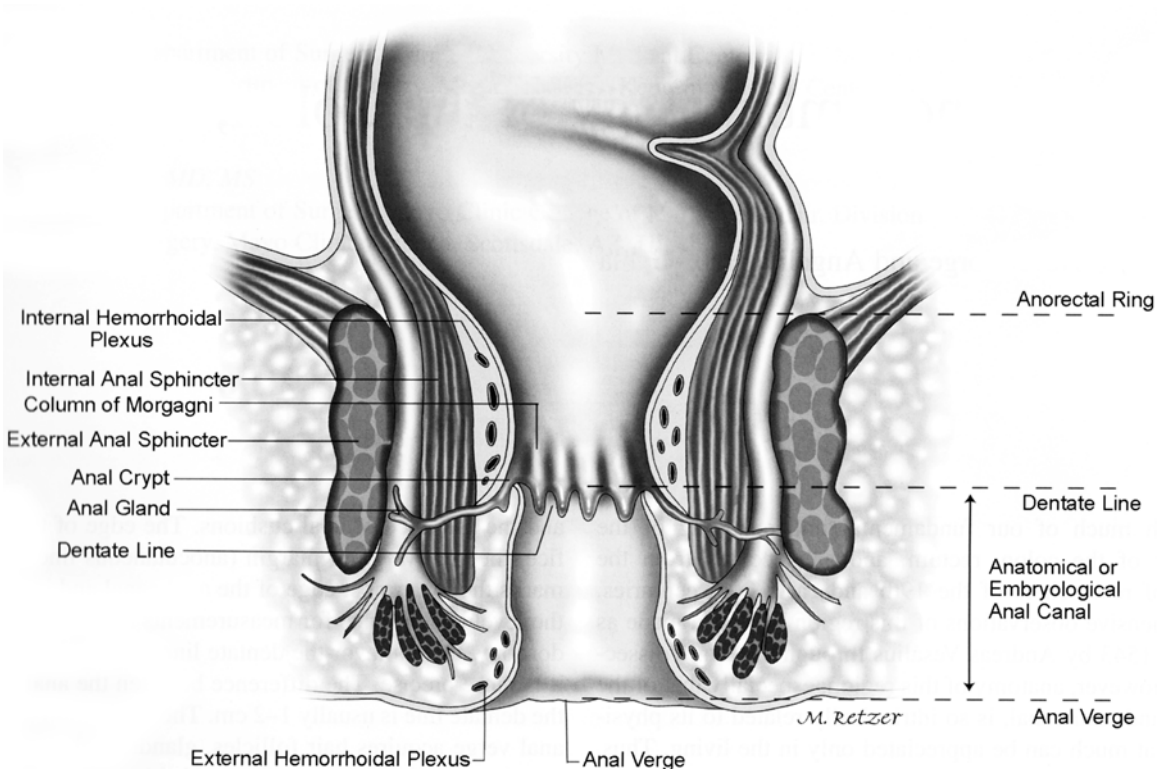

FIG. 49.1 Anatomy of the anal canal (Reprinted with permission from Jorge JMN, Habr-Gama A. Anatomy and Embryology of the Colon, Rectum, and Anus. In: Wolf BG, Fleshman JW, Beck DE, Pemberton JH, Wexner SD (eds). The ASCRS Textbook of Colon and Rectal Surgery. Chapter 1. New York: Springer Science + Business Media, 2007: 1-22)

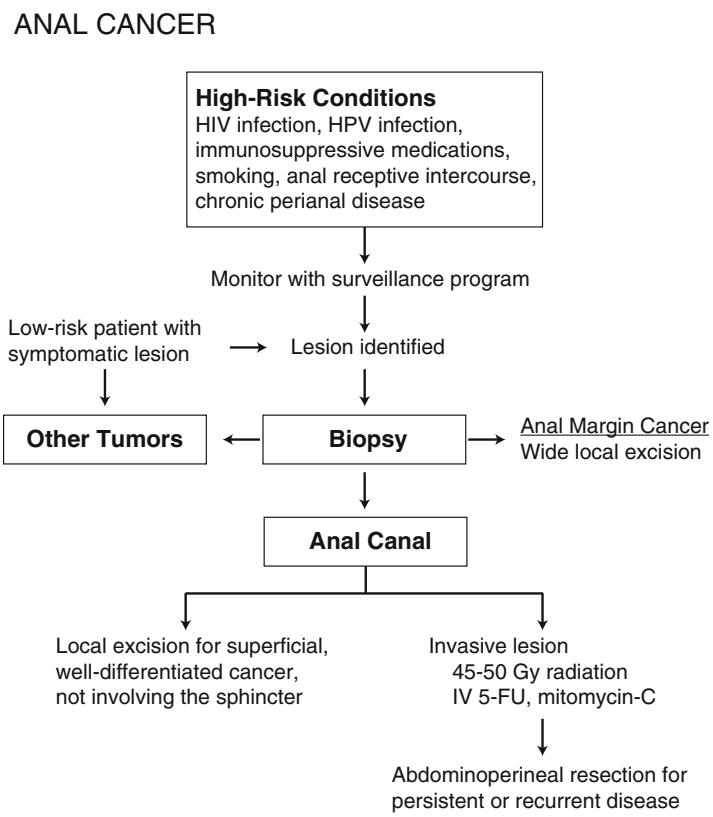

FIG. 49.2 Treatment algorithm for anal cancer. *HIV* human immunodeficiency virus, *HPV* human papilloma virus, *5-FU* 5-flourouracil

anesthesia in the majority of instances. Occasionally, an examination and biopsy under general anesthesia, in the operating room, is required if the patient is uncomfortable and experiencing pain. Once a cancer diagnosis is confirmed with a tissue biopsy, additional tests are needed to complete cancer staging such as CT scan or MRI of the abdomen and pelvis, chest x-ray, and anal ultrasound. Endoanal ultrasound may assist in determining the depth of tumor and assessment of nodal basins in anal canal cancers. All patients should be tested for HIV and undergo complete colonoscopy [1].

Treatment

Squamous Cell Carcinoma

Anal Margin

These lesions resemble those occurring elsewhere on the normal skin. Workup should include examination of the lymph node basins including physical examination of the inguinal lymph nodes. Superficial well-differentiated, small lesions (<2 cm and some advocate up to 4 cm) may be treated with wide local excision. An attempt should be made to get a 1 cm margin. Larger tumors (>4 cm) and poorly differentiated tumors should undergo chemoradiation first [1].

Anal Canal

Local excision is an adequate operation for well-differentiated cancers less than 2 cm that are limited to the mucosa and submucosa. A combined modality using chemotherapy and radiation is the initial treatment of choice for more advanced lesions. The most common regimen includes a radiation dose between 45 and 50 Gy, intravenous 5-flourouracil, and a single dose of mitomycin-C. Complete response rates of 85–90 % have been reported. After treatment has been completed, biopsies of the primary tumor may be done to check for persistent disease (Table 49.1) [2].

Management of Inguinal Lymph Nodes

Prophylactic groin dissections are not performed at any time, either at the initial diagnosis or when persistent or recurrent disease is detected. However, selective lymph node dissection can be done for local control of positive inguinal lymph nodes. If radiation is given, the radiated field should include any positive lymph node basins [2].

Advanced Disease

An abdominoperineal resection (APR) may be necessary for tumors that persist after chemotherapy and radiation or recur after such treatment. An APR is a radical operation that involves the removal of the rectum, rectal mesentery, anus, and the sphincter muscles with the creation of a permanent colostomy. Patients with recurrent anal canal disease should undergo an APR. An attempt can be made to re-excise recurrent cancers of the anal margin. Cisplatin or 5-flourouracil is used for palliation in patients with distant metastasis [1, 2].

TABLE 49.1. Staging of anal canal cancers.

Tumor status	T1: <2 cm T2: 2–5 cm T3: >5 cm T4: deep invasion			
Nodal status	N0: no LN N1: perirectal LN N2: unilateral iliac/inguinal LN N3: bilateral iliac/inguinal LN or N1 + N2			
Metastatic status	M0: no mets M1: distant mets			
Stage 0	Tis	N0	M0	
Stage1	T1	N0	M0	
Stage 2	T2,3	N0	M0	
Stage 3[a]	Any T	Any N	M0	(or T4, N0, M0)
Stage 4	Any T	Any N	M1	

[a]Note: Stage 3 is now divided into 3a and 3b

Other Perianal Neoplasms

Bowen's Disease

Bowen's disease is squamous cell carcinoma in situ or high-grade anal intraepithelial neoplasia (AIN). Bowen's disease may cause itching and may be associated with a raised lesion that in some instances encircles the anus. Fewer than 10 % progress to cancer. It is frequently found incidentally after an unrelated surgery (i.e., hemorrhoidectomy). Raised lesions should be excised. Broad-based or circumferential lesions are first managed with a mapping procedure whereby biopsies determine the extent of disease and subsequent surgery. Other treatment modalities include 5-flourouracil cream, imiquimod, photodynamic therapy, radiation, cautery ablation, and laser therapy [2].

Anal Adenocarcinoma

Anal adenocarcinoma arises from the mucosa of the transitional zone. It is indistinguishable from rectal adenocarcinoma. Adenocarcinoma in the anal region is often associated with chronic fistulas. Treatment includes neoadjuvant/adjuvant chemoradiation. Most will need an APR. However, wide local excision can be used for small, well-differentiated lesions that do not invade the sphincters [1, 2].

Melanoma

Perianal melanoma usually presents in an advanced stage and has a worse prognosis than other perianal tumors. Melanoma arises from the transitional epithelium of anal canal, anoderm, or the mucocutaneous junction. Due to its location, it can be confused with a thrombosed external hemorrhoid. Melanoma is treated either with wide local excision or APR. Indications for APR include tumors larger than 4 cm, bulky tumors, inability to achieve negative margins via local excision, and involvement of the sphincter muscle. Tumors greater than 10 mm are usually not curable. As with melanoma found in other parts of the body, some authors advocate that the patient with anal melanoma undergo sentinel lymph node mapping. Radiation and immunomodulators may improve local and regional control [1].

Basal Cell Carcinoma

Basal cell carcinoma can be seen in preexisting conditions such as basal cell nevus syndrome, xeroderma pigmentosum, immunodeficiency, radiation, chronic irritation, burn, and trauma. These tumors have variable clinical appearance (nodule, plaques, ulcers). Treatment includes wide local excision for tumors less than 2 cm. Larger lesions can undergo wide local excision with skin grafting or Mohs microsurgery. Larger lesions extending into the anal canal may need chemoradiation or APR, but this is extremely rare [1].

Paget's Disease

Paget's is an intraepithelial adenocarcinoma that arises from the apocrine glands and spreads into the overlying dermis. It may mimic a rash. Treatment includes a wide local excision with microscopically clear margins for noninvasive tumors. Positive margins require a re-excision. APR or adjuvant therapy is used if invasive cancer is found. Radiation is an alternative for poor surgical candidates [1, 2].

Lymphoma

Lymphoma is the second most common AIDs-related neoplasm. The anorectal area is devoid of lymphoid tissue, but can be "acquired" in chronic infections and anal receptive intercourse. Patients may have local (pain, pruritis, and drainage) or constitutional symptoms (fever, night sweats, and weight loss). Treatment includes chemotherapy (cyclophosphamide, doxorubicin, oncovin, and prednisone or CHOP) and radiation [1].

References

1. Welton ML, Varma MG. Anal Cancer. In: Wolf BG, Fleshman JW, Beck DE, Pemberton JH, Wexner SD (eds). The ASCRS Textbook of Colon and Rectal Surgery. Chapter 35. New York: Springer Science + Business Media, 2007: 482-500.
2. Nivatvongs S. Perianal and Anal Canal Neoplasms. In: Gordon PH, Nivatvongs S, eds. The Principles and Practice of Surgery for the Colon, Rectum, and Anus. Third ed. Chapter 18. CRC Press, 2007: 369-390.

50
Anorectal Pain

Leandro Feo and Nadav Dujovny

History and Physical Exam

Patients often seek medical advice for anorectal complaints. Symptoms are typically attributed to hemorrhoids, but in reality, there are numerous conditions that could cause pain or discomfort. Moreover, many patients tend to delay evaluation because of embarrassment. Most anorectal conditions are benign and easily treated. However, because of this inherent delay, many of these conditions often present as advanced disease necessitating more extensive treatment. Key historical points include whether anorectal pain is constant or intermittent and whether it is related to bowel movements (Fig. 50.1).

If the patient has obstructive symptoms or changes in bowel habits, the entire colon should be evaluated by colonoscopy. Anorectal assessment consists of inspection and palpation. The physician should be vigilant for anorectal cancer, and an anoscopic and/or proctoscopic evaluation should be considered in all patients. The patient can be positioned in the left lateral decubitus or in the prone jackknife position for the examination.

Intermittent Anal Pain

Anal Fissure

Anal fissure is one of the most common anorectal problems and is defined as a tear of the anoderm distal to the dentate line. Patients afflicted with this disorder commonly present with severe pain during and after defecation, which can last from a few minutes to several hours. Patients also present with the passage of bright red rectal blood. Anal fissures are caused by spasm of the internal sphincter, producing a lack of blood supply to the posterior commissure of the anal canal. As a result of this, wounds in the anoderm are ischemic, chronic, and nonhealing. A spastic sphincter typically precludes digital rectal exam or anoscopy.

An anal fissure may be classified as acute or chronic. **Acute** fissures are characterized by distinct mucosal edges and granulation tissue while **chronic** fissures have indurated edges, a lack of granulation tissue, a sentinel skin tag, hypertrophied anal papilla, and some degree of anal stenosis. Oftentimes, the internal sphincter muscle fibers are visible at the base of a chronic fissure. Ninety percent of anal fissures are located in the posterior midline, and the remainder are typically located in the anterior midline. Anterior fissures are more commonly seen in young women and are paradoxically related to low internal sphincter pressures. There is a subset of fissures that can be found in patients with inflammatory bowel disease, tuberculosis, syphilis, and immunosuppression such as HIV and cancer. Anal fissures within this subset are frequently located laterally and may present as multiple lesions.

Most acute anal fissures respond to general treatment measures such as sitz baths, stool softeners, increased oral fluid intake, and a high fiber diet. Treatment of chronic anal fissures is directed toward decreasing internal sphincter spasm and decreasing the elevated resting anal pressure. Chemical sphincterotomy with calcium channel blockers, topical nitroglycerin, or botulinum toxin injection is the initial approach. For those patients with recurrent fissures or when medical treatment has failed, surgery is advised. The internal sphincter is incised up to the dentate line through a lateral incision. Gross incontinence is rare afterward. In patients with anterior midline fissures and low resting anal pressures, fissurectomy and advancement flap are the treatments of choice if medical management has not been successful.

Levator Ani Syndrome (LAS)

Chronic proctalgia is characterized by rectal pain lasting at least 20 min in the absence of systemic or organic cause. Levator ani syndrome (LAS) is an intermittent, deep-seated rectal pain, which can resolve spontaneously. Currently there

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_50, © Springer New York 2015

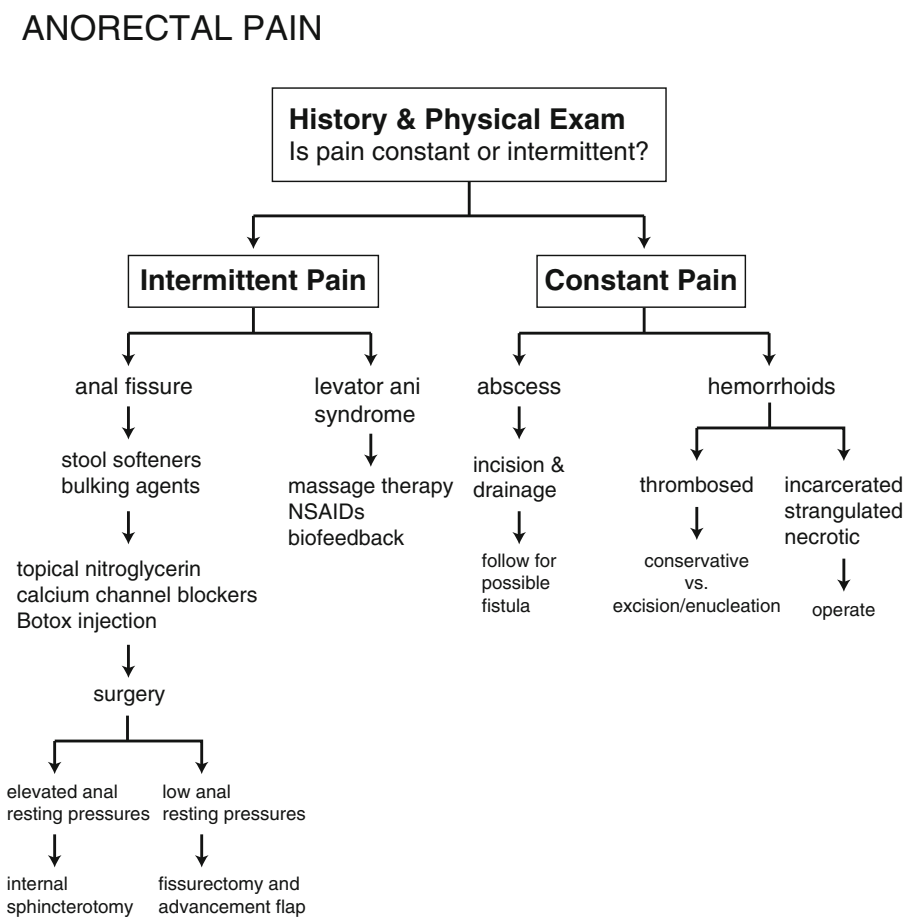

FIG. 50.1 Treatment algorithm for anorectal pain

is no consensus about the etiology of this condition; however, spasm of the pelvic floor muscles is a constant finding. Digital rectal examination should be performed in a patient complaining of chronic proctalgia; if the pain is elicited after palpation of the levator ani muscle, the patient is likely to have this condition. Despite this finding, LAS remains a diagnosis of exclusion – diagnosed after more common conditions have been ruled out clinically, endoscopically, and radiographically. At the present time there is no universal successful treatment in the management of this entity, the most common modalities include biofeedback, transrectal or transvaginal pelvic floor massage, and nonsteroidal anti-inflammatory agents with variable degrees of pain relief.

Constant Rectal Pain

Anorectal Infection

Anal abscesses and fistulas should be considered the acute and chronic phase of the same anorectal infection. Abscesses begin as an infection of the anal glands and crypts located at the dentate line, spreading to the adjacent space and structures. Abscesses are classified based on their location as perianal, ischiorectal, intersphincteric, or supralevator. Patient symptoms may vary based on abscess location. Perianal and ischiorectal abscesses produce localized pain associated with fever, erythema, tenderness, and fluctuance and are usually visible to the examiner. Intersphincteric and supralevator abscesses have deeper and more poorly localized pain and are often not visible to the examiner. In the circumstance of a supralevator abscess, a computed tomography (CT) scan of the abdomen and pelvis becomes a very useful diagnostic tool. Other causes such as diverticulitis or inflammatory bowel disease can lead to the development of a supralevator abscess.

The management of anorectal abscess is incision and drainage. Frequently, drainage can be performed in the emergency room or office. On occasion, the patient needs to be taken to the operating room for deeper and more extensive abscesses accompanied by fever or leukocytosis. An incision is placed on the area of fluctuance closest to the anal canal. This is done to decrease the potential distance of a fistula, if it were to occur. A recurrent abscess at the same

location should raise the suspicion for a fistula-in-ano. These often present with chronic drainage and nonhealing of the surgical wound. Treatment is aimed at eradicating the fistula without compromising fecal continence.

Hemorrhoids

Hemorrhoidal disease refers to "the state of symptoms attributed to the vascular cushions present in the anal canal." These submucosal vessels are sinusoids, not veins, and can exhibit arterial bleeding. Hemorrhoids are classified as internal or external depending on their location in relation to the dentate line. Presenting symptoms include mucosal protrusion, bleeding, mucus discharge, and pain. Pain occurs with thrombosis, strangulation, and gangrene and it is usually constant in nature.

External hemorrhoids are covered with squamous epithelium and occur distal to the dentate line. They have somatic innervations that allow for pain sensation. Treatment consists of topical agents to relieve pain, decrease swelling, and improve hygiene. Patients may present with an acute thrombosis of their external hemorrhoid. Treatment typically consists of analgesics, sitz baths, and fiber supplementation. If the thrombosis is extremely painful, then it should be excised. Typically, this can be done in the office or the emergency room.

Internal hemorrhoids are proximal to the dentate line and are covered by columnar epithelium. This area does not have somatic innervations and is insensate. These hemorrhoids tend to bleed and prolapse. Treatment starts with a high fiber diet and fiber supplementation. Smaller ones can be treated with rubber band ligation. Symptomatic larger ones require operative intervention, such as excision.

51
Rectal Bleeding

Jennifer Jolley

Overview

This chapter will focus on minimal hematochezia or bright red blood per rectum (BRBPR) in patients who are hemodynamically stable and not hemorrhaging. Patients may present with complaints of minimal amounts of red blood on toilet paper after wiping or having noticed a few drops of blood in the toilet bowl after defecation. According to some studies, approximately 15 % of the population experiences BRBPR, although this may be underreported especially in younger people. During the history and physical it is critical to delineate the type of bleeding and how much the patients have in order to help determine the etiology. The source may be from common anorectal pathology such as hemorrhoids, anal fissures, rectal ulcers, or proctitis; or it may be caused by sources higher in the colon such as polyps, diverticulosis, angiodysplasia, or neoplastic lesions.

Clinical Assessment

During the assessment of a patient with rectal bleeding, it is important to differentiate whether their history is consistent with minimal BRBPR, suggesting a lesion near the anal canal; or melena or maroon stool mixed with bright red blood, which may imply an upper GI, small intestine, or more proximal colonic pathology. Typically, blood that is seen only on the toilet paper indicates distal pathology while blood that has a different color (usually darker) is indicative of a more proximal source. In an infant or adult, rectal bleeding associated with pain and defecation may denote an anal fissure. Patients may have bleeding secondary to a thrombosed hemorrhoid that is demonstrated by a constantly painful lump unrelated to defecation. Chronic infection, inflammation, or malignancy may be suggested by the presence of systemic symptoms such as weight loss, fevers, or night sweats. A change in the frequency or caliber of stool may also support an underlying malignancy, while loose consistency of the stool is an indication that colitis may be present. In addition to considering the current presentation of symptoms, it is important to determine if the patient has other significant risk factors for cancer such as age, family history of colonic polyps or cancers, or history of inflammatory bowel disease (IBD). All of the aforementioned information should be taken into consideration when determining the clinical approach to identifying the etiology of rectal bleeding (Fig. 51.1).

During the physical exam, one must always carefully inspect the external anus and perform a digital rectal examination. Hemorrhoids or rectal prolapse may become more apparent by asking the patient to "bear down" during the exam. One may confirm the presence of anemia with laboratory testing, but a positive fecal occult blood test (FOBT) alone will help support the need for a thorough gastrointestinal evaluation. FOBT is an important part of the annual exam of patients, but it must be performed in a specific manner in order to be accurate. This test measures peroxidase activity that is present in both hemoglobin and certain foods (red meat, turnips, salmon, melons, horseradish, sardines, and vitamin C). These foods should be avoided for at least 48 h prior to the collection of the first stool specimen. Other recommendations include the cessation of aspirin or other nonsteroidal anti-inflammatory drugs for 7 days prior to and during the test period. Immunochemical FOBT tests that are now available tend to be more specific and not require any dietary restrictions. Because of these other factors that can affect the FOBT results, a negative FOBT does not preclude the patient from undergoing other tests.

Anoscopy or proctoscopy should be performed during the office visit in order to detect anorectal pathology. Anoscopy offers the best means to evaluate lesions in the anal canal and perform anal procedures to treat these lesions. Proctoscopy is able to assess the distal 20–25 cm of the colon and thus may be advantageous over anoscopy in order to see more possible pathology in the rectum and distal sigmoid colon.

Additional diagnostic testing with a sigmoidoscopy or colonoscopy is warranted for patients who have risk factors for cancer or IBD, those who have not had a colonoscopy,

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_51, © Springer New York 2015

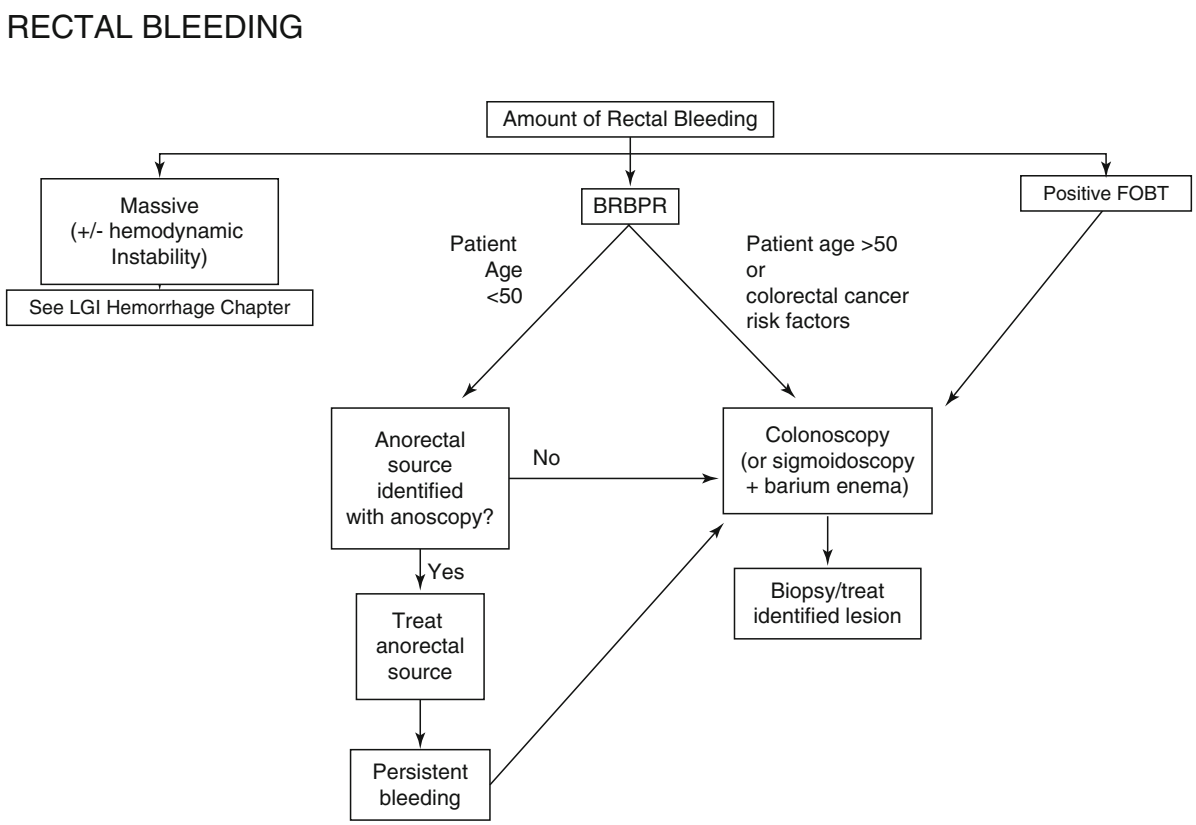

FIG. 51.1 Treatment algorithm for rectal bleeding. *BRBPR* bright red blood per rectum, *FOBT* fecal occult blood test

those with persistent BRBPR despite treatment, or those in whom there was no identifiable anorectal pathology to explain the BRBPR. Sigmoidoscopy only evaluates the distal 60 cm of the colon but does not require sedation and may be more available than colonoscopy. Some physicians support the use of sigmoidoscopy as a screening tool alone or in combination with double-contrast barium enema. However, many GI consensus statements recommend colonoscopy for all patients in whom an anorectal source of bleeding is not found and in patients who are older than age 50 even if there is a potential anal lesion. There is debate as to whether or not sigmoidoscopy is cost effective, as some studies have shown that up to 90 % of patients undergoing sigmoidoscopy eventually required further testing with colonoscopy whether the test was normal or abnormal. It is reasonable to consider flexible sigmoidoscopy as the initial diagnostic tool in patients with unexplained bleeding up to the age of 50; however, patients above age 50 should undergo colonoscopy regardless of whether or not anorectal pathology has been identified. Another possible diagnostic test, barium enema, is typically only reserved for patients whose colonoscopy was incomplete or inadequate.

Common Causes

As stated previously, various types of colonic or anorectal pathology can be the cause of BRBPR in patients. However, here we will focus on hemorrhoids, which are one of the most common causes of rectal bleeding, especially in patients less than 50 years old. Hemorrhoids occur as a result of engorgement of three fibrovascular cushions located in the anal canal in the left lateral, right posterior, and right anterior positions. They can be classified into internal or external hemorrhoids, the former occurring above the dentate line and the latter below the dentate line. Internal hemorrhoids are further classified into grades I through IV based on whether or not they prolapse and/or spontaneously reduce or are irreducible. Patients tend to have a history of chronic straining with constipation or prolonged attempts at defecation. Hemorrhoidal bleeding results from rupture of the internal hemorrhoids, which are supplied by the superior and middle hemorrhoidal arteries. It is rarely the cause of extensive lower GI bleeding and tends to be either noticed on the toilet paper after the patient wipes or seen dripping into the bowl. The bleeding may be associated with

hemorrhoidal prolapse and swelling but rarely with pain unless it is a grade IV internal hemorrhoid that is incarcerated and irreducible. On the other hand, thrombosed external hemorrhoids can be extremely painful. The different treatment options, medical versus surgical, vary for these two pathologies depending on the amount of symptoms, grade of the internal hemorrhoid, and duration of pain associated with the external hemorrhoid. Medical therapy includes increasing hydration, fiber supplementation, stool softeners, topical ointments, and sitz baths. Surgical intervention includes rubber band ligation, sclerotherapy, coagulation, or operative hemorrhoidectomy.

52
Cholelithiasis

Michael Tran and Daniel J. Deziel

Introduction

Cholelithiasis is a common condition. Management of the patient with gallstones depends on the symptomatic nature of the condition and on relevant nonbiliary considerations.

Asymptomatic Cholelithiasis

The majority of individuals with cholelithiasis have few or no related symptoms; however, 2 % of the asymptomatic population will develop biliary symptoms each year. *Prophylactic or anticipatory cholecystectomy is not necessary for most gallstone patients provided they are truly asymptomatic*. The clinician should evaluate all abdominal symptoms, since not all patients present with typical complaints of right upper quadrant pain (Fig. 52.1).

There are circumstances that may be exceptions to the above and for which cholecystectomy for asymptomatic gallstones should be considered on an individual basis:

1. Patients undergoing an unrelated major open abdominal operation. Cholecystectomy can usually be performed efficiently and safely in this setting and will prevent postoperative cholecystitis that can occur.
2. Patients with anticipated long-term parenteral nutrition because they develop gallstones and sludge. Morbidity, mortality, and the need for emergency biliary operations are more frequent in these patients.
3. Women anticipating pregnancy. Biliary stasis associated with pregnancy may increase the risk of developing symptoms. Symptomatic, and especially complicated cholelithiasis during pregnancy, can endanger both the mother and the fetus.
4. Immunosuppressed organ transplant patients may develop serious septic biliary complications. Some centers routinely screen potential kidney recipients for gallstones and recommend cholecystectomy prior to transplantation if stones are found. However, this remains a debated topic, and a strategy of expectant management for asymptomatic patients is recommended by others.
5. Children, because of their higher lifetime risk of becoming symptomatic.
6. Individuals with known gallstones who will be without access to surgical care for prolonged periods of time.
7. Individuals (especially younger) who belong to populations at high risk for gallbladder cancer.

Current evidence does not support diabetes mellitus alone as an indication for cholecystectomy for patients with asymptomatic gallstones. However, experience indicates that diabetic patients who develop complications of cholelithiasis or require emergency biliary operations have a higher morbidity and mortality than do nondiabetic individuals.

Symptomatic Cholelithiasis

Most patients who develop symptoms will continue to experience them and are at risk of complications unless the gallbladder is removed.

Acute Cholecystitis

Acute cholecystitis occurs in 10–20 % of individuals with symptomatic gallstones. The typical presentation includes right upper quadrant abdominal pain and tenderness, Murphy's sign, nausea, vomiting, fever, and leukocytosis. *Elevations of liver enzymes and amylase are not unusual even without common bile duct stones or pancreatitis*. Ultrasound confirms gallstones and may demonstrate other characteristic features, such as gallbladder distention, thickening of the gallbladder wall, pericholecystic fluid, and a "sonographic Murphy's sign." Radioisotope scanning with iminodiacetic acid derivatives will fail to visualize the gallbladder but is not usually necessary. Initial medical management includes intravenous fluid and electrolyte resuscitation, intravenous antibiotics, and pain control.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_52, © Springer New York 2015

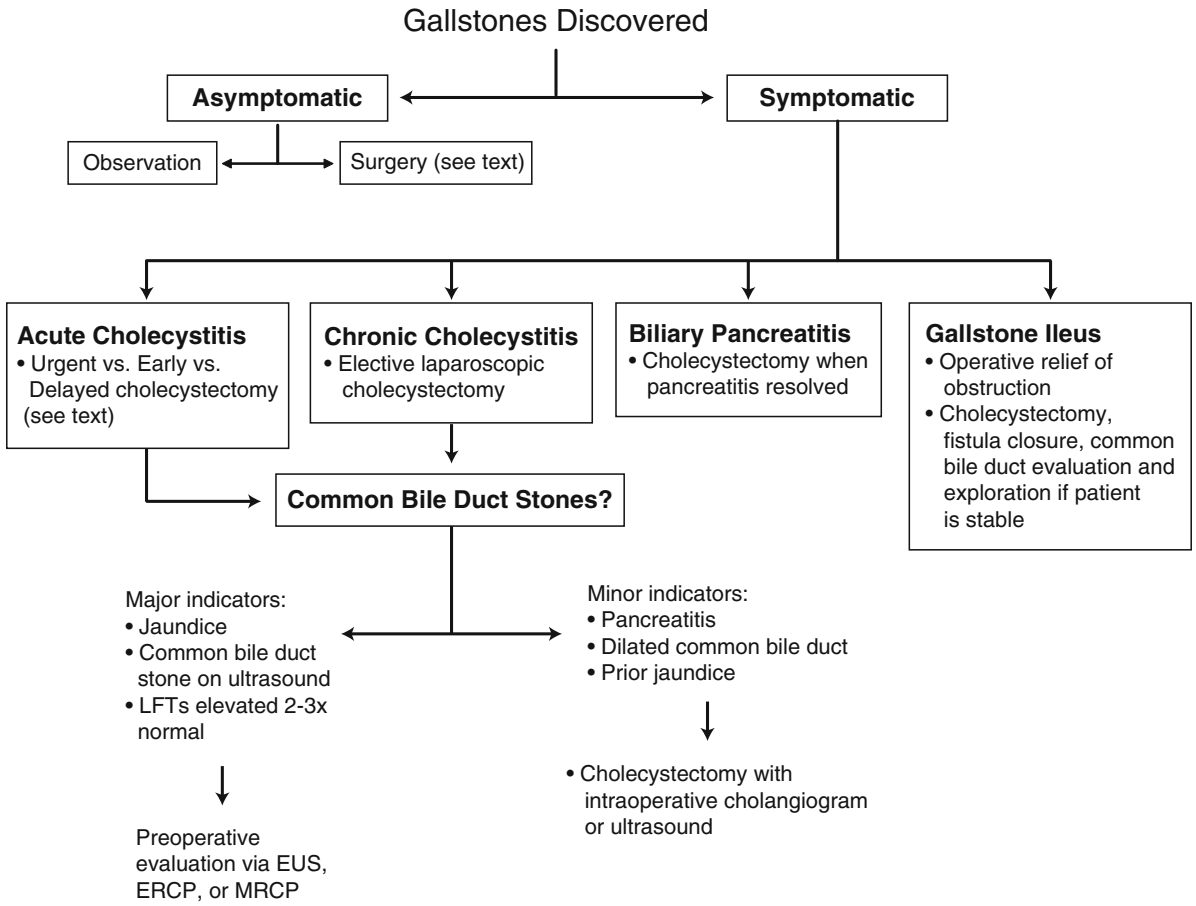

Fig. 52.1 Treatment algorithm for cholelithiasis. *LFTs* liver function tests, *EUS* endoscopic ultrasound, *ERCP* endoscopic retrograde cholangiopancreatography, *MRCP* magnetic resonance cholangiopancreatography

Ten percent of patients with acute cholecystitis warrant *urgent cholecystectomy* because of toxic manifestations. This applies particularly to patients with diabetes or significant vascular disease who may progress rapidly to gangrene and who do not tolerate sepsis well. Operation can usually be performed after several hours of intravenous hydration and correction of metabolic abnormalities. *Early cholecystectomy* means that the patient is admitted to the hospital, treated medically (antibiotics, hydration), and operated upon generally within 48 h. This is preferred for most patients with acute cholecystitis because it can be performed safely (and usually laparoscopically) and leads to the quickest resolution of the problem. *Delayed cholecystectomy* is performed days to weeks after diagnosis. This may be indicated for patients with serious comorbid medical conditions that can be improved during the interim or for patients who refuse initial operation. However, delayed cholecystectomy has been associated with higher rates of open conversion and postoperative complications as well as increased hospitalization times and cost. Moreover, one-third of medically treated patients do not improve or worsen during their initial hospitalization. Percutaneous cholecystostomy can be a temporizing measure for the initial nonoperative management of patients with acute cholecystitis who are a prohibitive operative risk.

Common Duct Stones

Approximately 10 % of patients operated on for gallstone disease will have stones in the common bile duct at some time during their course. *Choledocholithiasis is as frequent in patients with acute cholecystitis as in patients with chronic cholecystitis*. A reasonable probability (>50 %) of choledocholithiasis exists in patients with (1) obstructive jaundice, (2) a common bile duct stone seen on transabdominal ultrasound (note, however, that transabdominal ultrasound is not a sensitive test for identifying common bile duct stones), and possibly (3) serum alkaline phosphatase levels elevated greater than two to three times normal. Under these circumstances, preoperative evaluation of the bile duct by either endoscopic ultrasound (EUS) or magnetic resonance cholangiopancreatography (MRCP) is appropriate. If choledocholithiasis is found, then endoscopic stone extraction is usually performed prior to cholecystectomy.

Less specific indicators of choledocholithiasis include pancreatitis, mild elevations of liver function tests, a dilated common bile duct on abdominal ultrasound, or a history of prior jaundice. Patients with these findings can usually proceed directly to laparoscopic cholecystectomy and

intraoperative imaging of the bile duct consisting of intraoperative cholangiography and/or laparoscopic ultrasonography. Patients who have had preoperative endoscopic retrograde cholangiopancreatography (ERCP) should be considered for intraoperative cholangiography because of the possibility of missed or retained bile duct stones following ERCP. Patients selected for preoperative ERCP are at higher risk for common bile duct stones at the outset, and the incidence of stones found during intraoperative evaluation is typically higher than it is for patients without preoperative ERCP.

Most surgeons perform intraoperative imaging with cholangiography, which is most accurate when done with fluoroscopy. Intraoperative ultrasonography is highly accurate for the detection of common bile duct stones and is more sensitive than cholangiography but is not as widely practiced.

If intraoperative imaging does not reveal common bile duct stones, then cholecystectomy is completed. If common bile duct stones are identified, they should be removed at the time of surgery when feasible. Often this can be accomplished laparoscopically. If the laparoscopic approach is not successful or possible, then traditional open common bile duct exploration may be necessary, providing the common bile duct is of sufficient caliber. Intraoperative ERCP with endoscopic sphincterotomy can be considered in some circumstances but is more logistically complex and less frequently undertaken. If the stones are small or questionable and nonobstructing, then the operation may be completed without intraoperative manipulation of the common duct. In this situation, it may be advisable to leave a transcystic catheter and external drain. Clinically significant retained stones are generally managed by endoscopic extraction postoperatively.

Patients who are septic merit special consideration. If the clinical picture suggests acute cholecystitis, prompt operation is indicated even if choledocholithiasis is also suspected. If the clinical picture suggests cholangitis as the primary problem, initial treatment is medical. Urgent bile duct decompression is indicated for patients with toxic cholangitis (hypotension, obtundation) who do not respond promptly to fluid resuscitation and antibiotics. This represents 5 % of patients with cholangitis. Urgent decompression is usually accomplished endoscopically but can also be achieved by operation or by placement of percutaneous transhepatic catheters.

Chronic Cholecystitis

Chronic cholecystitis can be diagnosed on the basis of symptoms of episodic postprandial right upper quadrant and epigastric pain; gallstones may be identified by ultrasonography. If major indicators of common bile duct stones are present, then the common bile duct evaluation is performed preoperatively as previously discussed.

Patients with chronic cholecystitis and no indicators of choledocholithiasis are treated by elective laparoscopic cholecystectomy. Intraoperative imaging of the bile duct is selective according to surgeon preference.

Biliary Pancreatitis

The diagnosis of biliary pancreatitis is usually made in patients with abdominal pain and gallstones, accompanied by hyperamylasemia/hyperlipasemia. However, not all patients with gallstones and elevated serum amylase or lipase have pancreatitis. Pancreatic enzymes can be elevated with acute cholecystitis or choledocholithiasis in the absence of pancreatic inflammation. Nonetheless, when biliary pancreatitis is suspected, the initial management is medical. The majority of patients with biliary pancreatitis have *mild* pancreatitis. Cholecystectomy is performed when the patient's symptoms have improved (usually 24–72 h). Preoperative ERCP is *not* necessary unless other more reliable indicators of choledocholithiasis, such as jaundice or cholangitis, exist. In patients with *severe* biliary pancreatitis, delayed cholecystectomy is carried out after medical management and resolution of the pancreatitis.

Gallstone Ileus

Gallstone ileus is intestinal obstruction caused by a sizeable gallstone that has entered the intestines through a fistula. The most common location of the fistula is between the gallbladder and the duodenum (cholecystoduodenal) and the most common site of obstruction is in the small intestine (ileum). A preoperative diagnosis can be made on the basis of clinical suspicion (elderly female patient, intestinal obstruction, no prior abdominal surgery) and radiologic detection of air in the biliary tract (pneumobilia). The primary goal of treatment is to relieve the intestinal obstruction. This is accomplished by removal of the intestinal stone(s) after the patient has been properly resuscitated. Cholecystectomy, fistula closure, and evaluation of the common bile duct is carried out for patients who are stable and in whom the right upper quadrant dissection is not deemed unduly hazardous. Extraction of the obstructing gallstone should be performed through an enterotomy in non-inflamed bowel. It is important to evaluate the remainder of the bowel during surgery because additional gallstones can be found about 10 % of the time.

53
Jaundice

Amina Merchant and Harry Richter

Introduction

Jaundice is a common clinical sign characterized most recognizably by yellow staining of the conjunctiva, mucous membranes, and skin due to an elevated bilirubin level in the blood. The majority of bilirubin (about 4 mg per kg produced daily) derives from the breakdown of red blood cells and their constituent hemoglobin. The remainder results from ineffective erythropoiesis and the degradation of muscle and cytochromes. In health, circulating unconjugated bilirubin is taken up by hepatocytes, where it becomes conjugated with glucuronic acid. Conjugated bilirubin is then excreted from the liver through bile, stool, and urine.

Jaundice may represent predominately excess unconjugated bilirubin ("indirect bilirubin") or an excess of unconjugated and conjugated ("direct") bilirubin. Clinical measurement of serum bilirubin includes "fractionation" of the total concentration into these two moieties and provides the first and critical clue to diagnosis (Fig. 53.1).

Unconjugated Hyperbilirubinemia

Elevated unconjugated bilirubin can be due to increased bilirubin production, decreased hepatocyte uptake, or impaired intrahepatic conjugation. Increased production is attributable to acute or chronic hemolysis or to resorption of the heme component of a hematoma. A CBC with peripheral smear, reticulocyte count, and hemoglobin electrophoresis can distinguish whether hemolysis is due to spherocytosis or sickle cell disease. A hematoma may form spontaneously (often because of excessive anticoagulation) or from trauma. Careful review of medications that can cause hemolysis or bleeding is essential. Aggressive transfusion of red blood cells will also contribute to an overload of bilirubin as banked cells lyse.

Impaired uptake or conjugation of bilirubin can occur from a variety of medical reasons, including congestive heart failure, portosystemic shunts, several medications, and a number of specific syndromes, such as Gilbert's. Unconjugated hyperbilirubinemia is rarely pronounced (i.e., <5 mg/dl) unless liver function is also compromised.

Mixed Conjugated and Unconjugated Hyperbilirubinemia

Pure conjugated hyperbilirubinemia is exceedingly rare; all common causes of conjugated hyperbilirubinemia in fact also result in some degree to unconjugated hyperbilirubinemia as well. The next diagnostic distinction to be made is whether the cause is mechanical obstruction of the biliary tract or else an acute or chronic liver dysfunction, for example, hepatitis. Clinical history (e.g., risk factors for viral hepatitis, symptoms of gall stones) and physical exam (e.g., stigmata of liver disease, or a palpable gallbladder) form the basis of a provisional diagnosis. Measurement of serum concentration of liver enzymes ALT, AST, and alkaline phosphatase, commonly but inaccurately called liver function tests ("LFTs"), provides a reasonably sensitive discrimination between obstructive and hepatic causes. Marked (8–10X normal or greater) elevation of aminotransferase levels with normal or near-normal alkaline phosphatase strongly suggests primary liver disease. Conversely, marked elevation of alkaline phosphatase with near-normal aminotransferases points to biliary obstruction. Moreover, greater magnitude elevations of alkaline phosphatase are found with fixed, high-grade biliary strictures (mainly malignant) while smaller elevations are usually seen with the less complete or permanent obstruction caused by common bile duct stones.

Obstructive Jaundice

A preliminary impression of obstructive jaundice needs confirmation and subsequent identification of the anatomic level and, if possible, the cause of the biliary obstruction. Routine

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_53, © Springer New York 2015

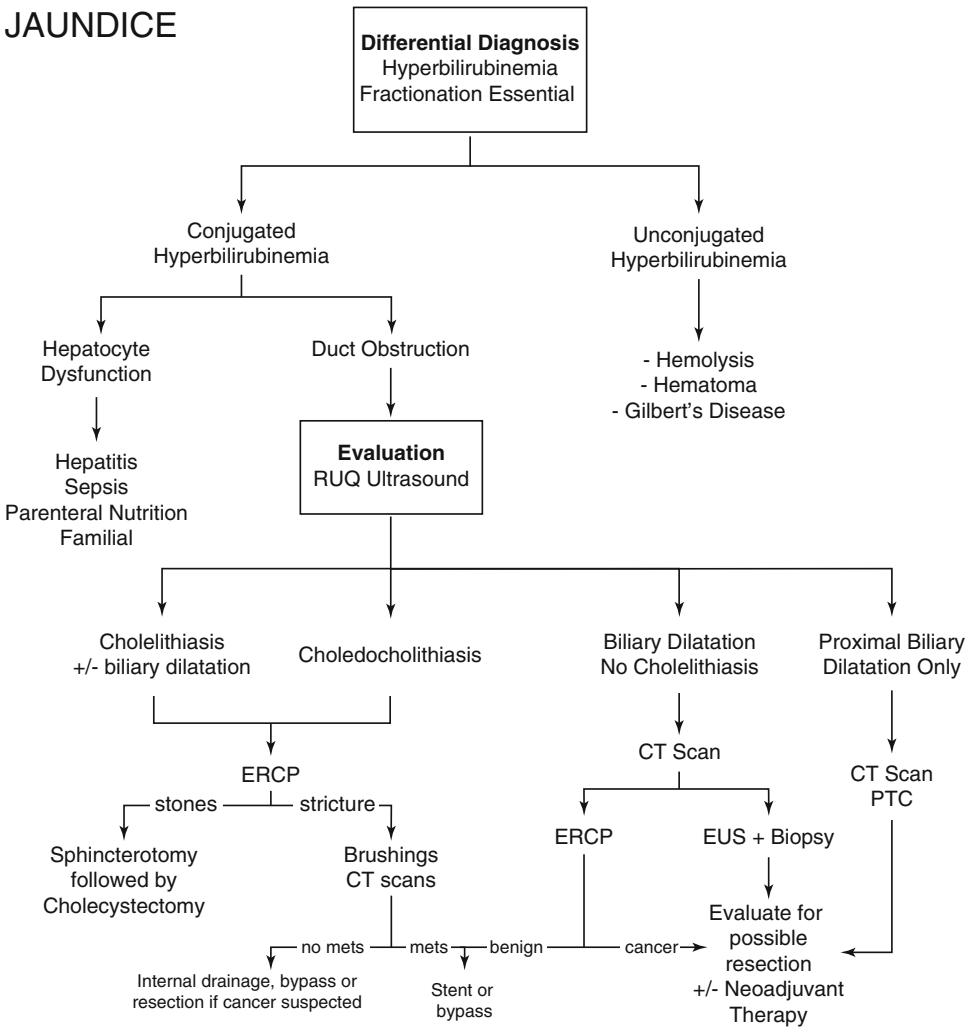

FIG. 53.1 Treatment algorithm for jaundice. *RUQ* right upper quadrant, *ERCP* endoscopic cholangiography, *CT* computed tomography, *PTC* percutaneous transhepatic cholangiography, *EUS* endoscopic ultrasound

right-upper-quadrant ultrasound provides a sensitive, harmless, and relatively inexpensive screen. Expect to identify (when present) dilated intra- and/or extrahepatic bile ducts, a distended gallbladder, gallbladder (but not common duct) stones, and any sizeable pancreatic head mass or pseudocyst. Furthermore, liver cirrhosis, steatosis, and ascites are usually apparent. Addition of Doppler blood flow measurement may diagnose hepatic veno-occlusive disease.

When ultrasound discloses only cholelithiasis and a dilated bile duct in a generally healthy patient, the diagnosis of choledocholithiasis is warranted, and endoscopic cholangiography (ERCP) with clearance of stones from the common bile duct prior to cholecystectomy is appropriate. Endoscopic ultrasound (EUS) and magnetic resonance cholangiography (MRCP) are also highly sensitive for diagnosing common bile duct stones.

Obstructive jaundice not due to stones is caused by neoplastic, inflammatory, or posttraumatic stricture. Malignant or benign neoplasms are most common, including tumors of the head or uncinate process of the pancreas, the ampulla of Vater or the periampullary duodenum, or the bile duct itself. Chronic pancreatitis, pancreatic pseudocyst, and autoimmune pancreatitis may cause biliary stricture. Traumatic stricture usually follows duct injury during cholecystectomy but may occur during gastric or liver resection or other procedures.

Evaluation of non-calculous obstruction of the bile duct includes cross-sectional imaging – either CT scan or MRI scan, including coronal and sagittal image reconstruction. Mass lesions suggestive of neoplasm are often sampled by EUS-guided fine needle aspiration (cytology) or biopsy. A positive tissue diagnosis and characterization of malignancy

enables neoadjuvant therapy when indicated and eliminates the need to establish a tissue diagnosis intraoperatively. Obstruction at the level of the right and left hepatic duct confluence (e.g., Klatskin tumor) likely demands percutaneous transhepatic cholangiography (PTC) to precisely delineate the proximal extent of disease.

Obstructive jaundice may selectively be improved preoperatively by endobiliary stent placement during ERCP (at which time cytological specimens are obtained). Reasons to do so include very deep jaundice, concomitant cholangitis, and need to delay operation to improve comorbid conditions.

54
Liver Mass

Marc G. Mesleh

Introduction

Due to the increasing utilization and sensitivity of radiologic imaging, the discovery of liver masses has risen dramatically, and approximately 250,000 patients are diagnosed with a liver lesion annually in the United States. Multiple imaging modalities are available that provide a wealth of information when interpreted correctly. As the morbidity and mortality associated with hepatic surgery continues to decrease, the clinical decisions surrounding these lesions have become more complex.

Symptoms

The majority of liver masses are asymptomatic and found incidentally (Fig. 54.1). When present, the most common symptom associated with liver masses is abdominal pain due to stretching of Glissen's capsule. Patients may present with jaundice from an underlying liver pathology or biliary obstruction. Early satiety is caused by large masses creating gastric outlet obstruction. Finally, peritonitis can result from spontaneous rupture, and DIC can result from a consumptive coagulopathy secondary to hemorrhage.

History and Physical Exam

Risk factors for liver masses include cirrhosis, chronic hepatitis, oral contraceptives, anabolic steroids, and carcinogens such as aflatoxin, Thorotrast, and vinyl chloride. Physical examination should focus on detecting stigmata of cirrhosis, such as ascites, abdominal wall varices, and telangiectasias. Importantly, the liver is a frequent site of metastatic disease due to its anatomic and physiologic properties. Therefore, a thorough medical history and physical exam are paramount in determining if the hepatic mass is a metastatic deposit from a distant primary tumor. Blood work should focus on liver function panels and tumor markers such as AFP, CEA, and CA 19-9.

Imaging Studies

Clinicians are armed with a vast array of diagnostic tests that have unique capabilities. Ultrasound is the least invasive test and is the easiest way to differentiate solid from cystic masses. Intraoperative ultrasound is even more sensitive at detecting smaller lesions. Computed tomography (CT) scans are becoming increasingly sensitive and are most helpful when intravenous (IV) contrast is administered. Magnetic resonance imaging (MRI) technology continues to develop, and many liver lesions have characteristic appearance with this modality. Sulfur colloid scans are nuclear medicine tests, which detect uptake by Kupffer cells. Finally, positron-emission tomography (PET) scans are increasing in popularity for metastatic disease and are performed in conjunction with CT scans at many institutions.

Tissue diagnosis can be obtained percutaneously via image-guided fine needle aspiration (FNA) or core needle biopsy. The currently accepted indications for biopsy include inconclusive imaging studies and a patient history of previous malignancy. These tests are not benign and have a measurable risk of intraperitoneal hemorrhage, hemobilia, pneumothorax, infection, and bile leak.

Solid Lesions

Peripheral Enhancing Lesions: Hemangioma

Hemangiomas are the most common benign tumor of the liver and overall prevalence has been estimated to be 5–20 %. These lesions are more common in women by a ratio of 6:1. Greater than 50 % are asymptomatic at diagnosis, but they may cause bleeding or disseminated intravascular coagulation, known as Kasabach-Merritt syndrome. CT scan with IV contrast reveals a well-circumscribed hyperattenuating lesion with lobular margins and a sequential filling of contrast

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_54, © Springer New York 2015

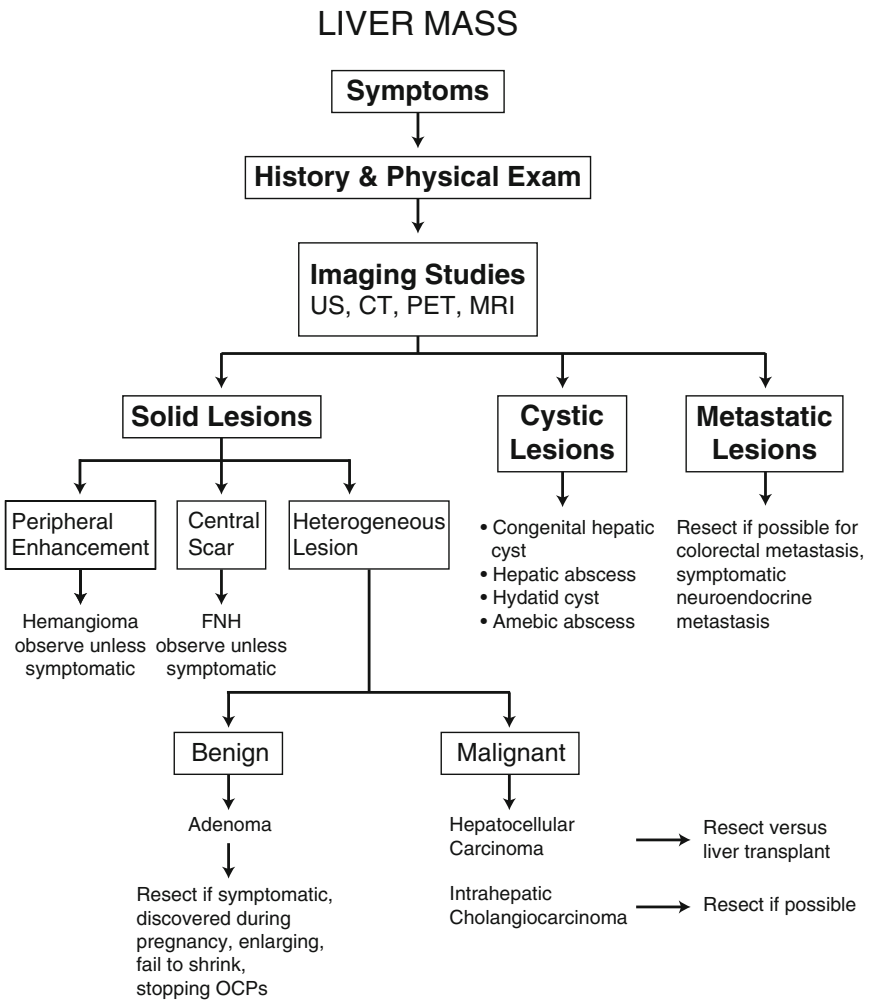

FIG. 54.1 Treatment algorithm for liver mass. *CT* computed tomography, *PET* positron-emission tomography, *MRI* magnetic resonance imaging, *FNA* fine needle aspiration, *OCP* oral contraceptive

from the periphery of the lesion toward the center and delayed washout. MRI reveals a well-defined lobular mass that is hypointense on T1-weighted images and hyperintense on T2, with characteristic peripheral nodular enhancement with gadolinium. Imaging for hemangiomas is typically diagnostic, and **biopsy is strongly contraindicated due to the risk of bleeding**. While these lesions are typically managed with observation, resection is indicated for symptomatic tumors and those in whom imaging is inconclusive and malignancy cannot be ruled out.

Central Scar: Focal Nodular Hyperplasia

Focal nodular hyperplasia (FNH) is a common lesion (3 % prevalence in adults) and represents 8 % of benign liver lesions. The etiology is thought to be a congenital vascular malformation or a vascular injury that elicits a hyperplastic response from the liver parenchyma. Greater than 80 % of these lesions are asymptomatic at diagnosis, but large lesions may cause symptoms by their mass effect. CT scans reveal a hyperdense lesion on arterial phase with a central scar, which becomes markedly more hyperdense on delayed images. MRI is the preferred modality for diagnosis as this central scar is classically hyperintense on T2-weighted images. Biopsy of these lesions is rarely needed because imaging is diagnostic for the majority of cases. There is no risk for malignant transformation, and surgical resection is not indicated for these lesions unless symptomatic.

Heterogeneous Benign Lesion: Hepatic Adenoma

Hepatic adenomas are benign liver tumors that are composed solely of hepatocytes with no organization of portal triads. They are more prominent in women (9:1) and are strongly associated with oral contraceptive use in women and anabolic steroid use in men. Discontinuing these medications should be the first step in management. It should be noted

that their recent incidence has been declining due to reduced levels of estrogens in newer OCPs. While these lesions are often asymptomatic at diagnosis, 25–50 % may present with pain due to stretching of Glissen's capsule. Adenomas can also cause hemorrhagic shock due to spontaneous intraperitoneal rupture; this most commonly occurs when lesions are greater than 5 cm. CT scans show a heterogeneous lesion with a smooth border and intense contrast enhancement due to their hypervascularity. MRI with gadolinium shows strong enhancement during the arterial phase with very rapid washout in the venous phase. Imaging is typically diagnostic. Biopsy is discouraged due to the risk of seeding the tract. If performed, a biopsy of the adenoma should reveal only hepatocytes and no Kupffer cells. Adenomas have a low risk of malignant degeneration, which is estimated at 5–11 % for lesions greater than 5 cm. With the decreasing morbidity of hepatic surgery, the indications for resection have become broader and are no longer based simply on size. Resection is indicated for symptomatic lesions, adenomas discovered during pregnancy, lesions growing in size, and lesions that do not decrease in size with the cessation of OCPs.

Heterogeneous Malignant Lesions

Hepatocellular carcinoma (HCC) is an epithelial tumor arising from hepatocytes and constitutes 80 % of all primary liver cancers. The incidence is increasing and is greater than 4 per 100,000 persons. Risk factors include hepatitis B carriers, chronic hepatitis C infection, cirrhosis from multiple etiologies, hereditary hemochromatosis, and rarely environmental toxins such as aflatoxin. These lesions have a 3:1 male-to-female ratio. **Any lesion greater than 2 cm in a cirrhotic liver is HCC until proven otherwise**. AFP levels are elevated and can aid in the diagnosis of equivocal lesions. On CT scan, HCC shows enhancement on arterial phase with a peripheral washout on delayed imaging. Smaller lesions with concerning findings on CT scan can be biopsied to confirm the diagnosis.

Partial hepatectomy is the treatment for resectable lesions in patients *without cirrhosis*. Unfortunately only 5 % of HCC occurs in the absence of cirrhosis, the presence of which increases the operative mortality from 3 % to 7–25 %. A normal bilirubin and no evidence of portal hypertension are the most clinically important factors associated with the lowest operative mortality. Partial hepatectomy has a 1-year survival of 55–90 % and a 5-year survival of 10–50 %; however, 5-year recurrence remains high at 60–100 %. **Orthotopic liver transplant is indicated for lesions discovered within cirrhotic livers**. Liver donors are a limited resource and therefore OLT should only be considered for patients with low risk of recurrence within the transplanted organ. The Milan criteria have set forth the indications for transplant: a single lesion less than 5 cm or three lesions less than 3 cm each, producing a 5-year survival of approximately 77 %. Alternative therapies exist for lesions that are not amenable to partial hepatectomy or liver transplantation. These include trans-arterial chemoembolization, radiofrequency ablation, and radio-embolization.

Cholangiocarcinomas are the second most common primary liver malignancy, responsible for 10–15 % of cases. They are classified into three groups: peripheral cholangiocarcinoma, cholangiocarcinoma arising within the left or right hepatic ducts, or hilar cholangiocarcinoma (known as Klatskin's tumors), arising at the junction of the left and right ducts. Risk factors include primary biliary cirrhosis, primary sclerosing cholangitis, choledochal cysts, hepatolithiasis, hepatitis B or C, and parasitic biliary infections such as *Clonorchis*. With its recent increasing prevalence, nonalcoholic steatohepatitis (NASH) is an increasingly important risk factor. These lesions are diagnosed similar to HCC, as a liver mass in the setting of right upper quadrant (RUQ) pain and weight loss. CT scan reveals a low-attenuating mass with irregular rim-like peripheral enhancement and peripheral intrahepatic biliary ductal dilation. Histologically, cholangiocarcinoma resembles an adenocarcinoma, and care must be taken to distinguish this from a hepatic metastasis from a primary tumor elsewhere in the gastrointestinal (GI) tract. An R0 resection to microscopically negative margins is a potentially curative treatment and should be considered in patients suitable to undergo hepatic resection.

Cystic Lesions

Hepatic cysts are typically benign, present in 2.5 % of the population, and are solitary in approximately 70 % of cases. They are usually asymptomatic and incidentally discovered but can cause abdominal pain when large. Ultrasonography is often diagnostic in identifying a fluid-filled lesion. The presence of septations within the cyst or mural nodularity raises the concern for cystadenoma or cystadenocarcinoma. CT scans reveal a non-enhancing fluid-filled lesion, and MRI shows a very high intensity on T2-weighted images due to the high water content. Biopsy of the cyst wall should only be performed when mural nodules raise the suspicion of malignancy. Symptomatic simple cysts can be treated with percutaneous drainage and sclerosis. Operative interventions include marsupialization or cystectomy performed via open or laparoscopic techniques.

Cystic lesions can also be infectious in etiology. Hydatid cysts arise from an infection caused by the tapeworm *Echinococcus granulosus*. CT scan shows a thick-walled cyst with calcifications and daughter cysts. These are diagnosed via serology. Treatment of these lesions is resection via peri-cystectomy after injecting the cyst with hypertonic saline or EtOH to prevent the anaphylaxis associated with spillage of cyst contents. Amebic liver abscess are caused by *Entamoeba histolytica* and present as single large cysts. Serology confirms diagnosis and these cysts resolve with

metronidazole. Pyogenic liver abscess typically arise from a distant infectious source and present with RUQ pain, fever, chills, and weight loss. Treatment consists of percutaneous drainage and 4–6 weeks of antibiotics.

Metastatic Lesions

Portal venous drainage from the GI tract flows through the liver and, therefore, it is a major source of metastases. Metastatic tumors of the liver are more common than primary hepatic malignancies in Western countries. The most common malignancies associated with hepatic metastasis include colorectal cancer, parasitic adenocarcinoma, breast cancer, gastrointestinal stromal tumors, lung cancer, melanomas, lymphomas, pancreatic islet cell tumors, renal cell carcinoma, and neuroendocrine tumors. These metastatic lesions are typically hypo-vascular, and therefore hypointense on contrast CT and MRI. The only exceptions are neuroendocrine and renal cell carcinoma metastasis, which are hypervascular lesions. The indicators for resection are growing, and now include colorectal cancer, breast cancer, symptomatic neuroendocrine metastasis, and some stromal tumors.

55
Portal Hypertension

Paul R. Balash and Edie Y. Chan

Introduction

Portal hypertension is defined as *increased pressure in the portal vein and/or its tributaries*. The main cause is obstruction along the path of venous blood flow from the portal vein, through the liver, into the hepatic veins. Portal hypertension is less commonly due to increased portal venous inflow. Portal pressure normally ranges from 6 to 8 mmHg and increases to greater than 12 mmHg in portal hypertension (can exceed 50–60 mmHg). *Cirrhosis is the most common cause of portal hypertension in the United States*, with the majority being due to excessive alcohol consumption. Other causes of portal hypertension include portal vein thrombosis (most common cause in children) and schistosomiasis (worldwide leading cause of pre-sinusoidal portal hypertension). Causes of portal hypertension are separated into prehepatic, intrahepatic, and posthepatic causes (Table 55.1). Collaterals and varices are formed by dilation of small, preexisting venules (Table 55.2), and these act to decompress the portal circulation into the systemic circulation.

Clinical Manifestations

Portal hypertension can lead to substantial complications, including ascites, variceal bleeding (esophageal, splenic, and gastric), hepatorenal syndrome, hepatopulmonary syndrome, hepatic encephalopathy (asterixis), and spontaneous bacterial peritonitis. Manifestations stem from portosystemic collateral formation, liver disease, and hyperdynamic circulation (Fig. 55.1, Table 55.3).

Diagnosis

Ascites is the accumulation of intraperitoneal serous fluid secondary to increased hydrostatic pressure and decreased colloid pressure caused by decreased protein production. The vast differential diagnosis necessitates paracentesis in patients with newly formed ascites. The ascites from cirrhotic patients is a transudative fluid with a predominance of monocytes and less than 250 neutrophil cells/mL. If fluid white blood cell count is greater than 500 cells/mL, with the majority being neutrophils, spontaneous bacterial peritonitis should be of concern. Functional hepatic reserve must also be assessed, along with portal venous anatomy and hepatic hemodynamics. The Child-Pugh classification is a method of defining risk associated with any intervention for portal hypertension and is the conventional method for assessing functional hepatic reserve (Table 55.4). A liver biopsy facilitates determining the cause of cirrhosis and the activity of the liver disease. The biopsy can be obtained percutaneously, or via a transjugular or laparoscopic approach if coagulopathy or moderate ascites dictate. Portal pressure can be indirectly measured via a hepatic venous wedge pressure or directly by transhepatic or umbilical venous cannulation of the portal venous system. Venous phase visceral angiography is now being supplanted by computed tomography (CT) angiography as a means of visualization of the portal venous anatomy and frequently performed prior to any portosystemic intervention. An alternative is duplex ultrasound, which can assess portal venous patency, portal blood flow, and portal shunt patency. When varices are of concern, endoscopy is the diagnostic gold standard.

Treatment

Treatment of portal hypertension is directed at the specific complication of the disease process (e.g., ascites, encephalopathy, and bleeding esophageal varices) and can be considered prophylactic or therapeutic.

Treatment of Ascites

Medical management is the mainstay of treatment for ascites caused by liver failure and is successful in 90 % of patients.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_55, © Springer New York 2015

TABLE 55.1. Causes of portal hypertension.

Prehepatic	Portal vein thrombosis
	Splenic vein thrombosis
	Congenital atresia
	Extrinsic compression
Intrahepatic	Cirrhosis
	Alcoholic
	Postnecrotic, hepatitis
	Biliary
	Hemochromatosis
	Wilson's disease
	α(alpha)1-antitrypsin
	Congenital hepatic fibrosis
	Schistosomiasis
	Hepatoportal sclerosis
	Idiopathic
Posthepatic	Budd-Chiari syndrome
	Veno-occlusive disease
	Graft versus host
	Chemotherapy
	Radiation
	Constrictive pericarditis

TABLE 55.2. Venous collaterals in portal hypertension.

Collateral	Decompression
Esophageal varices	Left gastric vein to azygos vein
Splenophrenic veins	Splenic and short gastric veins to phrenic veins
Caput medusae	Umbilical vein to epigastric veins
Veins of Sappey	Liver capsule to diaphragm
Veins of Retzius	Retroperitoneal veins to vena cava
Hemorrhoids	Inferior rectal vein to external iliac veins

Management begins with fluid and sodium intake restriction (1,000 mg/day) along with an aldosterone antagonist (25 mg BID) to promote fluid loss and sodium excretion. Furosemide (20 mg/day) is occasionally added to increase fluid loss. Therapeutic paracentesis is performed when the ascitic volume hinders respiration or mobility.

Treatment of Encephalopathy

Management of encephalopathy consists of protein intake restriction, increased ammonia excretion, and decreased ammonia production. Lactulose is commonly used as a cathartic, but it also alters the pH of the colon and inhibits bacterial production of ammonia. Intraluminal antibiotics, such as rifaximin, is used to decrease bacterial flora, thereby decreasing overall bacterial production of ammonia.

Treatment of Acute Bleeding

Resuscitation and stabilization should be the initial principal focus. Intravenous fluid should consist of D5W to prevent salt retention and worsening of ascites. A nasogastric tube should be placed to prevent aspiration. Several units of packed RBCs should be typed and crossed. A Foley catheter and central venous catheter should be placed to monitor fluid status. Coagulopathy should be corrected and platelets transfused for goal platelet count greater than 50,000/mm^3.

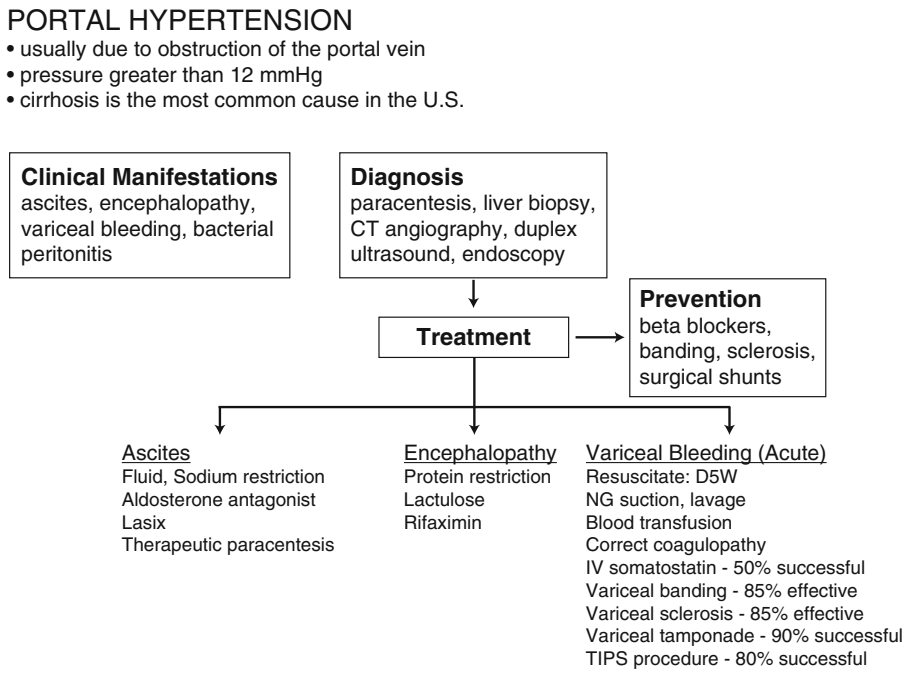

FIG. 55.1 Treatment algorithm for portal hypertension. *NG* nasogastric, *IV* intravenous, *TIPS* triangular intrahepatic portacaval shunt

TABLE 55.3. Clinical manifestations.

Signs of portosystemic collateral formation	Dilated veins in the anterior abdominal wall (umbilical epigastric vein shunts)
	Venous pattern on the flanks (portal-parietal peritoneal shunting)
	Caput medusa (tortuous collaterals around the umbilicus)
	Rectal hemorrhoids
	Ascites – shifting dullness and fluid wave (if significant amount of ascitic fluid is present)
	Paraumbilical hernia
Signs of liver disease	Ascites
	Jaundice
	Spider angiomas
	Gynecomastia
	Dupuytren contracture
	Muscle wasting
	Palmar erythema
	Asterixis
	Testicular atrophy
	Splenomegaly
Signs of hyperdynamic circulation	Bounding pulses
	Warm, well-perfused extremities
	Arterial hypotension

TABLE 55.4. Child-Pugh classification for hepatic functional reserve.

Measure	1 point	2 points	3 points
Bilirubin (mg/dL)	<2	2–3	>3
Albumin (g/dL)	>3.5	2.8–3.5	<2.8
INR	<1.8	1.8–2.3	>2.3
Ascites	Absent	Mild	Moderate
Encephalopathy (grade)	Absent	Grade 1–2	Grade 3–4

Class A, 5–6; class B, 7–9; class C, 10–15

Intravenous somatostatin, or its analogue octreotide, is used to increase splanchnic vasoconstriction and decrease portal venous flow and is successful in treating acute variceal bleeding in 50 % of patients. Vasopressin was used in the past to increase vasoconstriction along with nitric oxide to counteract the systemic side effects, though it has been shown to be less effective than somatostatin and is no longer used.

Gastric lavage should be performed with warm solution to evacuate the stomach prior to endoscopic evaluation in a stable patient. Endoscopic therapy is effective in approximately 85 % of patients and consists of variceal ligation (banding) and sclerosis. Banding is performed in a similar manner to hemorrhoidal banding. Sclerotherapy is performed by injecting 1–2 mL of sclerosant into each varix to cause edema and scarring and obliterate the variceal lumen. Gastric varices are not as amenable to endoscopic management compared to esophageal varices.

Luminal tamponade is another method of controlling variceal bleeding when pharmacologic and endoscopic treatments fail. When properly applied, variceal tamponade effectively controls bleeding in 90 % of cases. The Sengstaken-Blakemore tube has both a gastric balloon and esophageal balloon that can be inflated to apply pressure to bleeding varices. Inflation must be done under radiographic guidance with close monitoring of balloon pressure, only after an airway is secured. The use of the Blakemore tube or its alternatives (Minnesota or Linton tube) is only a temporizing measure and must be followed by definitive treatment as recurrent variceal hemorrhage often recurs after balloon deflation. Variceal tamponade can only be applied for 24–36 h due to risk of necrosis to underlying tissue, and care must be taken to not overinflate and cause perforation.

Transjugular intrahepatic portocaval shunt (TIPS) bypasses the diseased liver parenchyma and is effectively placed in 95 % of patients with cessation of bleeding in 80 % of patients with acute variceal bleeding. The jugular vein is cannulated and a wire is passed through the IVC into a hepatic vein. The hepatic vein wall is punctured with the wire, which then traverses the liver parenchyma before entering a large portal vein branch. A covered stent, which has replaced bare metal stents secondary to improved patency, is placed and the diameter is adjusted until cessation of variceal bleeding.

When variceal bleeding is not controlled by the aforementioned measures, a surgical portosystemic shunt is indicated. The portal venous system is decompressed by either partially or completely diverting flow via an end-to-side or side-to-side shunt into the systemic circulation (low-pressure inferior vena cava).

Prevention of Recurrent Bleeding/ Chronic Treatment

In stable patients without signs of variceal bleeding, prevention of recurrent bleeding can be accomplished by medical, endoscopic, radiologic, and surgical therapy. Medical therapy includes the use of B-blockers to decrease portal venous flow. Endoscopic therapy similar to what is used for acute bleeding (banding and sclerotherapy) can also be used to decrease recurrence. The goal is to eradicate all visible varices over a series of endoscopic interventions. Limitations of long-term endoscopic management are the possible increase in gastric variceal formation and a 30 % failure rate, though bleeding episodes are of decreased severity. TIPS, as described earlier, is also used to prevent recurrent bleeding and is increasing in popularity as compared to surgical portosystemic shunts. The two major complications associated with TIPS are increased rates of encephalopathy and thrombosis. Stenosis or occlusion is thought to be due to intimal hyperplasia along the covered stent and occurs in 20 % of patients by 1 year. As a result, there is still a chance of recurrent variceal bleeding and patients must be monitored every few months and treated with dilation or re-stenting as needed.

Surgical portosystemic shunts, though less commonly performed due to the advent of endoscopic therapy and TIPS, are still an option for chronic management of portal hypertension and variceal bleeding. Nonselective (total) portosystemic shunts completely divert portal blood flow away from the liver to the systemic circulation via direct anastomosis or interposition grafts (Table 55.5). The main complications of nonselective shunts are continued deterioration of hepatic function and hepatic encephalopathy. Selective portosystemic shunts are less likely to lead to hepatic encephalopathy by decompressing gastroesophageal varices while preserving portal flow to the liver. These shunts are as effective as nonselective shunts in preventing recurrent variceal bleeding, though they are contraindicated in patients with ascites.

TABLE 55.5. Nonselective (total) portosystemic shunts.

End to side	Portal vein (end) to vena cava (side)
Side to side	Portal vein (side) to vena cava (side)
Mesocaval	Superior mesenteric vein to vena cava
Central splenorenal	Splenectomy, splenic vein to renal vein

For patients with persistent variceal bleeding despite previous shunting or for those for whom shunting is contraindicated, non-shunt procedures may be considered. These operations halt the blood flow through varices and consist of either esophageal transection and reanastomosis or distal esophageal and stomach devascularization with splenectomy.

56
Carcinoid Disease

Faaiza T. Vaince

Introduction

Originally termed "Karzinoide" in reference to their indolent nature, carcinoid tumors are relatively slow-growing neuroendocrine neoplasms that secrete a diverse array of biologically active factors. The incidence in the United States is about 7–15 million a year and the overall 5-year survival is ~50 %.

Carcinoid tumors most commonly arise from enterochromaffin cells (Kulchitsky cells) in the gastrointestinal (GI) tract (75–87 %), specifically in the midgut (64 %), within 2–3 ft from the ileocecal valve. They can also be found in the respiratory tract (up to 28 %). Uncommon sites include the skin, retroperitoneum, ovary, prostate, or kidney (1–5 %). The bioactive factors they secrete vary with tumor location (Fig. 56.1). Foregut and respiratory tumors typically secrete serotonin and its precursor 5-hydroxytryptophan (5-HTP). Bronchial carcinoid tumors may also secrete ACTH or neuropeptides, while foregut GI tumors may secrete GI peptides and histamine. Midgut (jejunum to transverse colon) tumors produce serotonins and GI peptides but are less likely to produce 5-HTP. Hindgut tumors (left colon and rectum) may produce GI peptides but do not significantly produce any serotonin or 5-HTP.

Unfortunately, most carcinoid tumors are metastatic at presentation. Gastrointestinal carcinoids commonly present with bowel obstruction or vague abdominal pain that may be due to mucosal ulceration, intestinal obstruction, intussusception, adhesions, or hypermotility. Some intra-abdominal or retroperitoneal tumors may elicit a fibrotic desmoplastic reaction and may even cause a mass effect that impedes vascular flow resulting in mesenteric ischemia or venous congestion.

Pulmonary Carcinoids

Pulmonary carcinoid tumors comprise only 1–2 % of all lung tumors. Typical carcinoid pulmonary tumors have a low-grade malignant potential, whereas atypical carcinoids may act in a more aggressive fashion. These tumors tend to be centrally located in the main bronchi and may present with atelectasis, bronchial obstruction, hemoptysis, pneumonia, or pleural effusions. Many are asymptomatic and are discovered incidentally on a chest X-ray or computed tomography (CT) scan. Biopsy is required for diagnosis. Positron emission tomography (PET) scans and bronchoscopy are also utilized in the diagnosis.

Surgical resection with associated lymph node dissection remains the mainstay of treatment. Chemotherapy is of limited benefit but is still being studied in the form of molecular targeted therapy (i.e., angiogenesis inhibitors, tyrosine kinase inhibitors, and mTOR inhibitors). For patients with typical carcinoids that have undergone resection, the overall 5- and 10-year survival is 87–100 % and 87–93 %, respectively. Ten-year survival for atypical carcinoids is considerably lower at 30–50 %.

Gastric Carcinoids

Gastric carcinoids are amongst the rarest gastric malignancies (less than 0.5 %) but may comprise up to 10–30 % of all gastrointestinal carcinoids. They are broken into three types. **Type 1** gastric carcinoids are associated with chronic atrophic gastritis. **Type 2** gastric carcinoids are associated with Zollinger-Ellison syndrome/MEN type 1 syndrome. **Type 3** tumors are not associated with hypergastrinemia and have a higher malignant potential. Gastric carcinoids may present with abdominal pain, hematemesis, diarrhea, or gastric outlet obstruction but may also be incidentally discovered during endoscopy.

Treatment of gastric carcinoids is dictated by the size of the tumor. Smaller tumors (less than 1.5 cm) without evidence of local invasion are usually amenable to endoscopic excision with follow-up endoscopic surveillance. Intermediate lesions (1–2 cm) should be surgically excised while larger lesions (greater than 2 cm), which are usually type 3 lesions, warrant aggressive local resection, antrectomy, and local lymph node

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_56, © Springer New York 2015

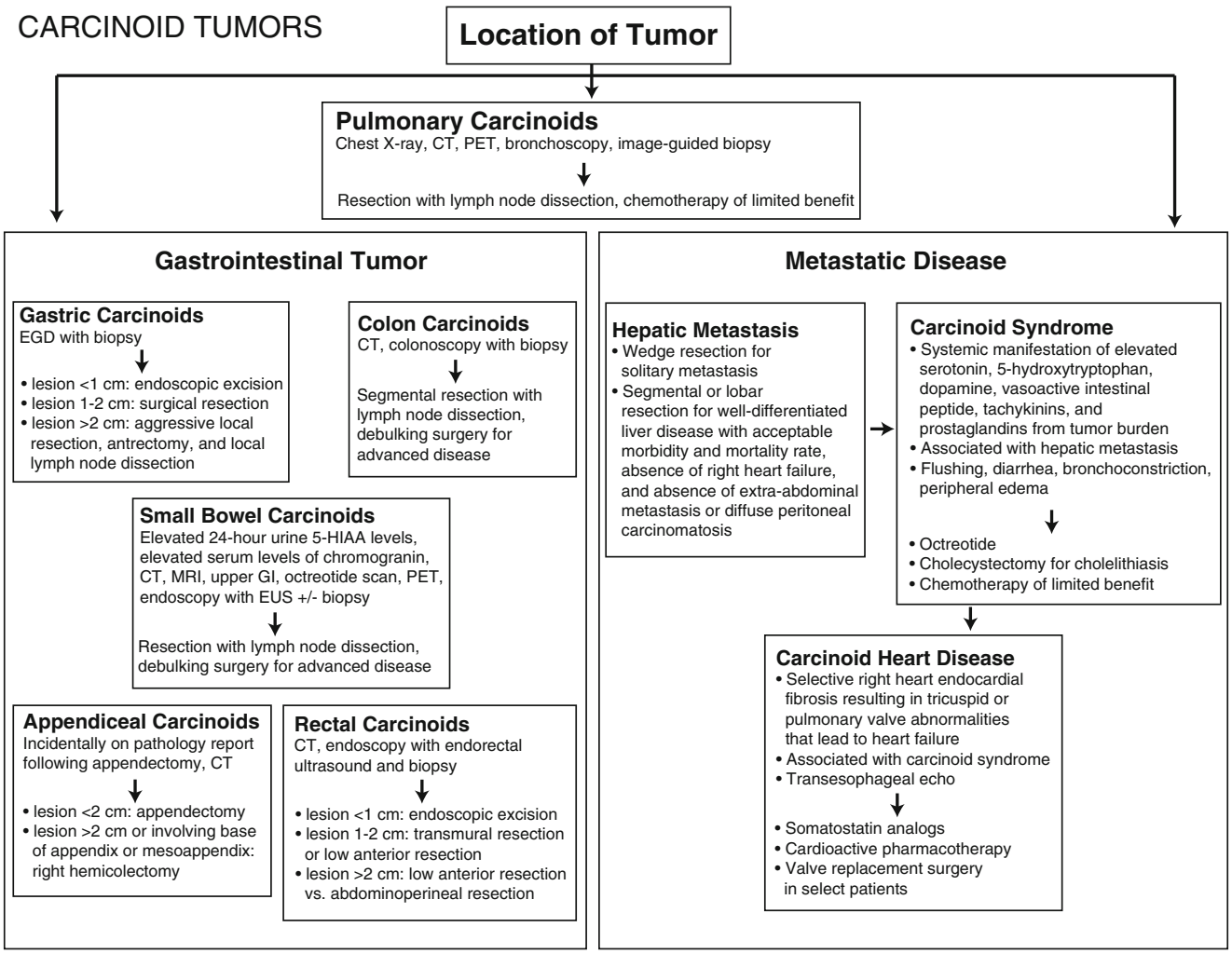

FIG. 56.1 Algorithm for diagnosis and treatment of carcinoid tumors. *CT* computed tomography, *PET* positron emission tomography, *MRI* magnetic resonance imaging, *GI* gastrointestinal, *EUS* endoscopic ultrasound

dissection. Occasionally, gastric carcinoids are associated with carcinoid syndrome, which can be treated with histamine antagonists. Proton pump inhibitors or H2 blockers are also utilized if the tumor is associated with hypergastrinemia.

Small Bowel Carcinoids

About 64 % of carcinoid tumors arise from the small bowel. These tumors comprise 30–35 % of all malignant small bowel tumors, and their incidence increased fourfold from 1985 to 2005. They are most commonly located in the ileum (45 %), followed by the duodenum (18 %) and jejunum (6 %).

Because of their indolent nature and submucosal location, these tumors are typically small and found incidentally. Larger tumors may present with bleeding or obstruction and are typically metastatic at presentation. In up to 40 % of cases, there are multiple primary tumors, and in 20 % of cases, the tumors are associated with noncarcinoid synchronous or metachronous tumors – most commonly adenocarcinomas of the colon. Rarely, they can also be associated with multiple endocrine neoplasia type 1.

Diagnosis of midgut carcinoid is suggested by elevated 24-h urine 5-HIAA levels. Elevated 5-HIAA (a serotonin metabolite) has a 73 % sensitivity for localized disease and 100 % specificity in predicting the presence of midgut carcinoid. Carcinoids also have elevated serum levels of chromogranin A (a glycoprotein secreted by tumor cells), though elevated levels can also be seen with proton pump inhibitors, atrophic gastritis, renal impairment, or inflammatory bowel disease. Thus its lack of specificity precludes its use as a diagnostic tool.

Contrast studies such as an upper GI, CT scan, or magnetic resonance imaging (MRI) may identify larger lesions or the presence of metastatic disease. Functional imaging studies, such as an octreotide scan or a PET scan, can also

aid in localizing these tumors. An octreotide scan has greater than 90 % sensitivity for detecting metastases. Endoscopy with ultrasound (EUS) may be utilized to identify duodenal lesions. In cases where the above studies have not successfully localized a suspected tumor, capsule endoscopy, double-balloon enteroscopy, and ileocolonoscopy can also be utilized.

Treatment of primary small bowel carcinoid tumors without evidence of metastases is resection with regional mesenteric lymphadenectomy. At the time of surgery, there should be a careful examination for additional lesions. Smaller duodenal lesions (less than 1 cm) without evidence of local invasion may be amenable to endoscopic resection. Debulking is indicated for locally invasive disease, even in the presence of hepatic metastasis, as it decreases incidence of obstructive and ischemic complications. For functional tumors, preoperative prophylaxis with octreotide is prudent. Intraoperatively, a carcinoid crisis can be treated with an intravenous (IV) bolus of octreotide followed by an octreotide infusion. Antihistamines, hydrocortisone, and albuterol are also used as warranted.

Postoperatively, patients should be followed every 3 months to evaluate for symptoms of recurrence and to obtain urinary 5-HIAA measurements and surveillance CT scans. The 5-year survival for limited and metastatic small bowel carcinoid disease is 65 % and 35 %, respectively.

Appendiceal Carcinoids

Though rare, appendiceal carcinoids are the most common tumors of the appendix and generally have a more favorable prognosis than other carcinoid tumors. They are most commonly discovered incidentally on pathology following an appendectomy. Ten to fifteen percent of appendiceal carcinoids are associated with a synchronous adenocarcinoma at another site. Therefore, postoperative imaging and endoscopic evaluation for additional lesions is warranted.

Appendiceal carcinoids are most commonly located at the tip (70 %) and at the time of discovery are typically less than 1 cm without evidence of metastases. Appendectomy alone is adequate for tumors that are less than 2 cm, located at the tip, and lack evidence of local invasion or lymph node metastasis. A right hemicolectomy is indicated for tumors that are greater than 2 cm in size, involve the base, or have evidence of local mesenteric or lymphatic invasion. The 5-year survival for limited local or regional disease is 85–95 % versus 34 % for tumors with distant metastases.

Colon Carcinoids

Colon carcinoids are rare, comprising only 8 % of all carcinoid tumors. They are most commonly located in the cecum and right colon and are also commonly associated with synchronous neoplasms (25–40 % of cases). Compared to small bowel tumors that may present with obstruction or bleeding, these tumors present with vague abdominal pain and anorexia or as an incidental finding on routine colonoscopy. Unfortunately, more than 60 % of tumors are metastatic by the time they present.

Regardless of the size of the lesion, segmental colon resection with regional lymphadenectomy and en bloc resection of additional involved tissues is the standard treatment for carcinoids of the colon. Debulking is indicated for unresectable tumors, as it significantly reduces symptoms and complications of advanced disease. The 5-year survival for localized and metastatic colonic carcinoids is 42 % and 71 %, respectively.

Rectal Carcinoids

Rectal carcinoids are more common than colon carcinoids. Fortunately they usually present early, have a lower malignant potential, and are rarely associated with carcinoid syndrome. They may present on digital rectal exam as firm, discrete, mobile submucosal lesions. Endoscopic biopsy usually establishes the diagnosis. Colonoscopy, endorectal ultrasound, and CT scans are utilized to evaluate extent of disease.

Similar to gastric carcinoids, treatment of rectal carcinoids is dictated by size. Smaller lesions that are less than 1 cm can be treated with endoscopic excision followed by endoscopic surveillance. Intermediate-size tumors that are 1–2 cm can be treated with transmural resection with adequate margins. Intraoperative pathology may be required to ensure disease-free margins. Larger lesions that are greater than 2 cm are usually associated with metastases and warrant aggressive surgical resection with a low anterior resection or abdominoperineal resection.

Metastatic Disease

Hepatic Involvement

More than 60 % of carcinoid tumors are metastatic at the time of diagnosis, and in most cases the liver is the involved organ. Wedge resection is indicated for solitary hepatic metastasis. Contraindications for liver resection include the presence of extra-abdominal metastases, diffuse peritoneal carcinomatosis, and significant comorbid illnesses. Those that undergo a curative resection have a 60–80 % 5-year survival rate. If surgical resection is not an option, other interventions such as ablative procedures, embolization, and liver transplantation may be considered. Functional liver metastases may also be palliated with debulking surgery.

Carcinoid Syndrome

Carcinoid syndrome is used to describe the clinical picture caused by the systemic release of a variety of humoral factors, including serotonin, 5-hydroxytryptophan, dopamine, vasoactive intestinal peptide, tachykinins, and prostaglandins. The syndrome occurs typically with GI carcinoid tumors that have metastasized to the liver or with tumors whose bioactive products do **not** undergo metabolism by the liver via the porta hepatis (e.g., retroperitoneal, ovarian, or pulmonary carcinoids). The syndrome can be seen in about 20 % of patients with midgut carcinoid tumors. It can be diagnosed by elevated levels of 5-hydroxyindoleacetic acid (5-HIAA) in the urine (>10 mg/24 h).

Classic symptoms of the syndrome include flushing (94 %), diarrhea (78 %), bronchoconstriction (19 %), and peripheral edema (19 %). Flushing is usually seen in the face, neck, and upper chest and can be transient or of longer duration. The diarrhea is usually episodic, explosive, and watery.

The symptoms of carcinoid syndrome, particularly the diarrhea and flushing, can be temporized with medical therapy, namely, long-acting somatostatin analogs such as octreotide and lanreotide. Long-term treatment with somatostatin analogs is associated with gallstone formation; accordingly, a cholecystectomy should be considered at the time of primary tumor resection. Chemotherapy has proven to be of limited benefit, though certain agents such as 5-fluorouracil, streptozocin, and interferon-alpha have been shown to decrease tumor burden.

Carcinoid Heart Disease

Cardiac involvement can be seen in up to 60 % of patients with carcinoid syndrome and is associated with an unfavorable clinical outcome. Specifically the syndrome is associated with selective right heart endocardial fibrosis that is initiated by the release of systemic serotonin, resulting in tricuspid or pulmonary valve abnormalities that lead to heart failure. The left heart is involved in 10 % of cases. Treatment includes medical management with somatostatin analogs and cardioactive pharmacotherapy. More recently, valve replacement surgery has increasingly been shown to improve clinical outcome and lengthen survival. Cytoreductive surgery and hepatic metastatic ablative therapies in the presence of carcinoid heart disease may also improve overall clinical outcome. Cardiac abnormalities should be screened for with an echocardiogram when carcinoid disease is diagnosed.

57
Inguinal Hernia

Lindsay Petersen and Keith W. Millikan

History

Approximately five million people in the United States have an inguinal hernia, and 800,000 repairs are performed annually (75,000 of these are recurrent hernias). The history is usually that of a groin bulge or pain occurring during straining or exercise. Approximately 25 % of men and 2 % of women during their lifetime will be diagnosed with an inguinal hernia. About 10 % of repairs are performed on women. Seventy percent of femoral hernia repairs are performed on women, but these are much less common than indirect or direct hernias, even among women.

Physical Examination

The physical exam is of *paramount* importance (Fig. 57.1). The most common physical finding is a *palpable* soft bulge produced by coughing or Valsalva's maneuver. More than 50 % of patients will have a *visible* asymmetry or bulge noted on inspection of both groins with the patient standing. Anatomically, inguinal hernias are classified as direct, indirect, or femoral (Table 57.1). Indirect and direct hernias cannot be differentiated on physical exam. Femoral hernias are palpable in the upper medial thigh at the outlet of the femoral canal.

Unilateral Hernia

The vast majority of inguinal hernias are reducible; the risk of incarceration or strangulation is approximately 1–2 % over a lifetime. Although uncommon, incarceration and strangulation are serious events that can lead to significant morbidity. Over time, inguinal hernias will increase in size, cause pain, become cosmetically unappealing, and be more difficult to repair. For these reasons, most surgeons agree that inguinal hernias should be repaired unless comorbid conditions preclude surgery. Almost all unilateral hernia repairs can be performed using local anesthesia and intravenous sedation. The most noteworthy complications concern the wound itself and include hematoma, seroma, and infection. Over the last decade, mesh repairs have become the gold standard. Tissue-to-tissue repairs are associated with longer recovery time and up to a 20 % lifetime recurrence rate. Mesh repairs have a recurrence rate of 1 %, are associated with less postoperative pain, and have faster recovery rates because of the tension-free nature of the repair. Use of biological mesh in inguinal hernia repair is mostly limited to repairs where there is concern for infection, since biologic mesh implants have decreased wound strength and are associated with increased rates of recurrence.

During the 1990s, laparoscopy emerged as a safe technique for inguinal hernia repair. Approximately 14 % of inguinal hernia repairs are now performed laparoscopically, but laparoscopic repair is associated with increased instrumentation cost and usually requires a general anesthetic (Table 57.2). In contrast, the mesh-plug hernioplasty, the Lichtenstein flat mesh, the Kugel preperitoneal patch, and the two-layer mesh repair can be performed under local or regional anesthesia (Table 57.3). A contraindication to preperitoneal mesh repair is prostate disease, such as cancer, benign prostatic hypertrophy, and previous prostatectomy. Onlay Lichtenstein repairs are preferred in the setting of prostate disease.

Incarcerated unilateral hernias require emergent repair; this is usually performed under general or spinal anesthesia. *The contents of the hernia sac must be examined* for possible ischemic intestine and a laparotomy or laparoscopy may be necessary if the bowel has retracted internally and viability is still a question. Because of the risk of infection, a permanent mesh should not be used in this situation.

Recurrent Hernia

For a recurrent hernia, it is important to determine whether a mesh or non-mesh repair was previously performed. For the latter, laparoscopic repair is the preferred approach to avoid

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_57, © Springer New York 2015

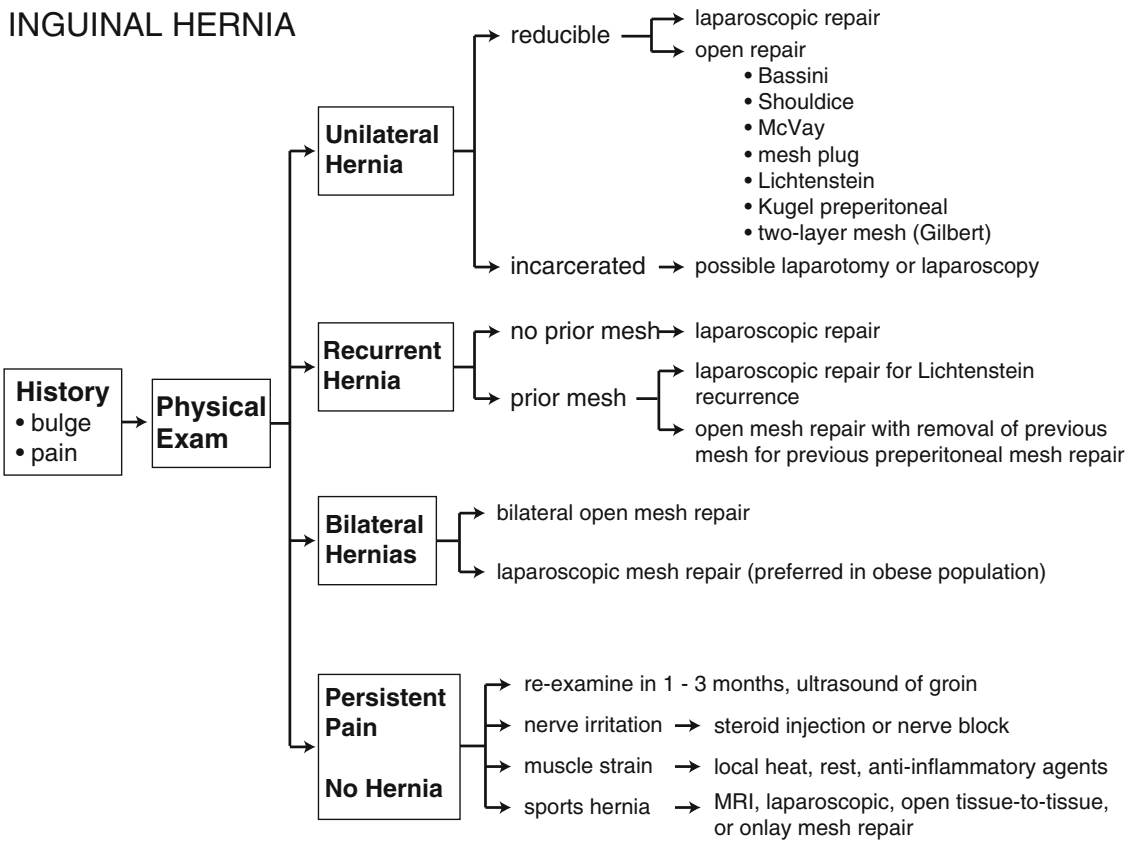

FIG. 57.1 Treatment algorithm for inguinal hernia. *MRI* magnetic resonance imaging

TABLE 57.1. Anatomy of inguinal hernias.

Type	Anatomy
Direct	Medial to inferior epigastric vessels Through Hesselbach's triangle
Indirect	Lateral to inferior epigastric vessels Through internal ring along the spermatic cord
Femoral	Below the inguinal ligament Through the femoral canal, medial to the femoral vein
Pantaloon	Combined direct and indirect inguinal hernias

TABLE 57.2. Types of laparoscopic repairs.

Type	Anesthetic	Technique
Transabdominal preperitoneal (TAPP)	General	The peritoneum incised from inside the abdomen and a mesh placed into the preperitoneal space and sutured to Cooper's ligament medially. The peritoneum then reapproximated over the mesh
Totally extraperitoneal (TEP)	General	The preperitoneal space accessed initially using a dissecting balloon, then a mesh placed in the preperitoneal space

scar tissue from the previous repair. Another possibility would be to perform a preperitoneal Kugel patch. This repair can also be performed using local anesthesia and intravenous sedation but has a steeper learning curve. For previously repaired mesh recurrences and multiple recurrent hernias, the preferred approach is a laparoscopic preperitoneal mesh repair if the preperitoneal space is free of mesh. In the cases where mesh was placed in the preperitoneal space, such as a preperitoneal Kugel patch, an open anterior approach under general anesthesia is preferred. The mesh should then be excised and a Lichtenstein or mesh-plug repair performed.

Bilateral Hernias

Ten percent of men with inguinal hernias also have contralateral hernias at presentation. Before mesh repairs became the gold standard, staged repairs were favored. However, since mesh insertion provides a tension-free repair, recurrence rates and speed of recovery are not compromised by a synchronous approach. Bilateral repairs can be performed by either an open mesh or laparoscopic preperitoneal repair. In the obese population, a laparoscopic approach would be preferred for bilateral hernias to limit the size of incisions and

TABLE 57.3. Types of open repairs.

Type	Anesthetic	Technique
Bassini tissue repair	Local with sedation	The internal oblique and transversus abdominis muscles and transversalis fascia secured to the inguinal ligament as a single layer with interrupted sutures
Shouldice tissue repair	Local with sedation	Similar to the above Bassini repair but done in layers with continuous suture to distribute tension evenly
Femoral (McVay)	Local with sedation	Mesh (or transversalis fascia if performing tissue repair) sutured to conjoined tendon and sutured below the Cooper's ligament medial to the femoral vein and to the inguinal ligament lateral to the femoral vein
Plug and patch	Local with sedation	Plug inserted into defect, only mesh covers the inguinal floor
Lichtenstein	Local with sedation	Flat mesh sutured to conjoined tendon, pubic tubercle, and inguinal ligament. Mesh split laterally to create a new internal ring
Kugel preperitoneal	Local with sedation	Large piece of flat mesh used to cover the entire inguinal floor in the preperitoneal space
Two-layer mesh (Gilbert)	Local with sedation	Two sheets of mesh with intervening connector. Underlay in the preperitoneal space with overlay along the floor of the inguinal canal

amount of tissue dissection. Many surgeons believe that laparoscopy offers quicker recovery and less morbidity for bilateral repair.

Persistent Pain

If a patient complains of groin pain, yet a hernia is not palpable, one should reexamine the patient in 3 months to make sure that a hernia has not been missed. Physical examination can be supplemented with an ultrasound examination while the patient is standing and coughing or straining. The differential diagnosis includes irritation of the ilioinguinal, iliohypogastric, or genital-femoral nerves. A steroid injection may be given and can be repeated every 2 weeks for three sessions; most likely the patient's nerve irritation will resolve. If pain persists, consultation with a pain center for phenol injections, radiofrequency ablation, or cryoprobe treatments are warranted. *The most common reason for groin pain with no palpable hernia is chronic muscle strain*. These injuries can take up to 4–6 months to heal. They are treated with local heat, the avoidance of exercise, and nonsteroidal anti-inflammatory agents. In most instances, these injuries will resolve. Groin exploration for patients with chronic injuries or nerve irritations will likely make the situation worse.

Other causes of groin pain without palpable hernia include pubic osteitis and sports hernia (athletic pubalgia). A sports hernia is a tear in the rectus abdominis or adductor longus as it inserts on the pubic bone, not a fascial defect. The main symptom is groin pain at the pubic tubercle with kicking or pushing off. Magnetic resonance imaging (MRI) is diagnostic for the condition. Laparoscopic hernia repair with mesh, tissue-to-tissue repair, and onlay mesh repairs with nerve dissections have all been performed to treat sports hernia. Pubic osteitis should be considered if consultation with a pain center has not been successful. The patient should receive a bone scan to rule out this condition.

Chronic pain following hernia repair lasting longer than 3 months after surgery can be classified into somatic and neuropathic categories. Somatic pain occurs with exertion or movement of the abdominal wall musculature and ligaments and is secondary to damage to these structures. Neuropathic pain is generally localized and sharp and usually indicates damage or entrapment of the ilioinguinal, iliohypogastric, or genital-femoral nerves. Neurectomies can be performed to treat neuropathic pain but should only be considered after evaluation by a pain specialist. Neurectomy is the last option for treatment, and variable success rates (50–90 %) are reported.

58
Abdominal Wall Defects

Julia Boll and Minh B. Luu

Introduction

The abdominal wall is an anatomically complex structure that serves the important role of supporting and protecting abdominal and retroperitoneal structures. Its essential muscular layers function toward this goal, as well as to enable twisting and flexing of the trunk. Abdominal wall or ventral hernias are defined as a protrusion of intra-abdominal or preperitoneal contents through the abdominal wall fascia and musculature. They are caused by weaknesses and defects in the abdominal wall and can be spontaneous from slow architectural deterioration of the muscular aponeuroses or acquired from previous surgical incisions. They are most easily categorized by their location on the abdominal wall (Table 58.1).

Anatomy

There are nine layers to the abdominal wall. The external oblique is the most superficial muscle of the lateral abdominal wall and its fibers run in an inferomedial direction. Deep to the external oblique is the internal oblique whose fibers run superolaterally. The deepest muscular layer of the abdominal wall is the transversus abdominis muscle, and its fibers course horizontally. Each of these muscles forms an aponeurosis that inserts into the linea alba, a midline structure demarcating the sides of the abdominal wall, and it is these aponeuroses that contribute to the anterior and posterior layers of the rectus sheath. On either side of the linea alba are the rectus abdominis muscles, which have fibers that run craniocaudally along the entire length of the anterior abdominal wall from the pubic symphysis to the xiphoid process.

Another important abdominal wall landmark is the arcuate line or line of Douglas. This line is 3–6 cm below the umbilicus, approximately at the level of the anterior superior iliac spines, and delineates the point below which the posterior rectus sheath is absent. Above the arcuate line, the anterior rectus sheath is composed of the external oblique aponeurosis and external lamina of the internal oblique aponeurosis, while the posterior rectus sheath has contributions from the internal lamina aponeurosis of the internal oblique as well as the aponeurosis of the transversus abdominis. Below the arcuate line, the external oblique, internal oblique, and transversus abdominis aponeuroses pass completely anterior to the rectus muscle, leaving no aponeurotic covering to the posterior rectus.

History and Physical

One should determine the duration, as well as the severity, of the patient's symptoms. It is important to rule out signs of strangulation and incarceration such as fever, skin warmth or erythema, nausea, emesis, constipation, and abdominal pain. Many times the symptoms are worsened with coughing or straining. Strangulation occurs more often in large hernias with small defects.

Physical exam will focus on the location of the hernia. Prior scars are important to take into consideration. During the exam, the anterior abdominal wall should be evaluated with the patient in both standing and supine positions. The edges of the fascial defect can be identified by palpation. The Valsalva maneuver can also help to demonstrate the site and size of a hernia.

Imaging and Preoperative Workup

Usually an abdominal wall hernia can be diagnosed clinically, without the use of imaging modalities. However, with increasing complexity of an abdominal wall hernia, it is important to define the extent and anatomy of the defect and to know if the bowel or other structures are involved (Fig. 58.1). Therefore, a proper diagnosis sometimes requires a computed tomography (CT) scan in addition to physical examination to visualize intra-abdominal contents within the hernia sac.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_58, © Springer New York 2015

TABLE 58.1. Types of abdominal wall hernias.

Type	Location	Strangulation	Characteristics	Repair
Rectus abdominis diastasis	Separation of rectus bellies at linea alba	None	Bulging of abdominal wall – mistaken for ventral hernia Midline aponeurosis intact with no actual hernia defect Can be congenital, acquired more common; occurs with age, obesity, or pregnancy	Diagnosed by exam, CT can differentiate from hernia and measure distance Correction with plication of midline aponeurosis
Umbilical – infant	Umbilical ring	Rare	Common and congenital; most common in premature or African American infants	Most close by age 2, frequently repaired if persist past age 5
Umbilical – adult	Umbilical ring	Rare, unless chronic ascites present	Acquired; more common in women or with ↑intra-abdominal pressure (i.e., obesity, pregnancy, ascites, abdominal distention)	Small/asymptomatic: follow clinically Symptomatic, large, incarcerated, or uncontrolled ascites: repair needed
Epigastric	Midline between xiphoid process and umbilicus	"Pain out of proportion" secondary to incarceration of preperitoneal fat	More common in men 3–5 % of population Usually small defects	Excise incarcerated preperitoneal tissue with simple closure of fascial defect; rarely big enough to require mesh
Spigelian	Through Spigelian fascia located at arcuate line Intraperitoneal, lies beneath ext. oblique aponeurosis	Relatively narrow necks increase risk for incarceration	Most are small and develop in adults Present with localized pain without a bulge	Always repaired -larger defects usually repaired using mesh
Obturator	Through the obturator canal formed by the pubic bone and ischium with covering obturator membrane	Weakening of membrane can result in hernia sac, incarceration, and strangulation	Howship-Romberg sign: medial thigh pain on internal rotation from compression of obturator nerve May present with complete or partial bowel obstruction	Posterior approach provides best access Open preperitoneal repair used if there's compromised bowel Repair membrane with sutures or small mesh
Grynfeltt's	Through superior lumbar triangle: 12th rib, paraspinal muscles, and internal oblique muscle	Lumbodorsal fascial weakness results in a sac that contains extraperitoneal fat – not prone to incarceration	Congenital or acquired More common than Petit's	Suture repair difficult because of immobile bony margins Repair usually completed with mesh secured to 12th rib and muscular borders
Petit's	Through inferior lumbar triangle: iliac crest, latissimus dorsi, and external oblique muscle	Lumbodorsal fascial weakness results in a sac that contains extraperitoneal fat – not prone to incarceration	Congenital or acquired	Suture repair difficult because of immobile bony margins Repair usually done with mesh secured to iliac crest, latissimus, and ext. oblique
Sciatic	Through greater sciatic foramen	Often how these hernias present; best diagnosed with ultrasound	Usually asymptomatic until complete intestinal obstruction; may present as uncomfortable or slowly enlarging gluteal mass or with sciatic nerve pain	Transperitoneal approach if bowel obstruction or strangulation. Transgluteal approach if diagnosis is certain and hernia is reducible
Perineal	Through pelvic floor – can contain fluid, fat, intestine, rectum, or bladder	Uncommon – symptom is usually a bulge protrusion worsened by sitting or standing	Mostly acquired after abdominoperineal resection or perineal prostatectomy without adequate pelvic floor reconstruction Primary hernias occur in older, multiparous women	Transabdominal or combined transabdominal/perineal approach Sac contents are reduced, small closed primarily, large repaired with mesh

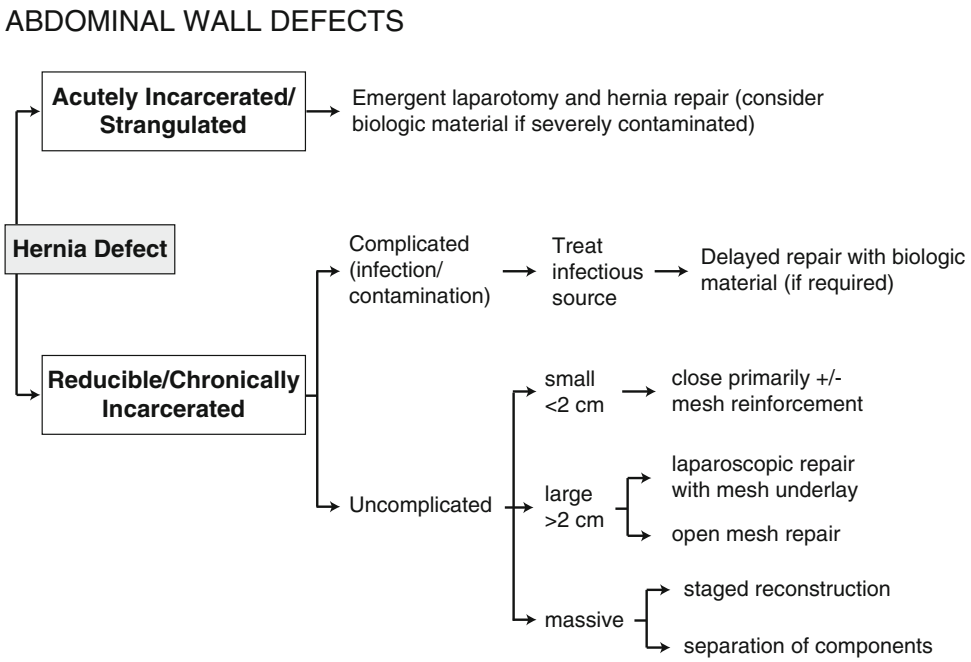

Fig. 58.1 Treatment algorithm for abdominal wall defects

Additionally, patients with comorbidities should undergo preoperative evaluation with chest X-ray, electrocardiogram (EKG), and/or pulmonary function tests to ensure that they have enough vital capacity to undergo a potentially large abdominal operation that may compromise respiratory function. Some patients present with grossly contaminated wounds, and it is vital to control the infection prior to definitive surgical repair, as this increases the likelihood of a successful abdominal wall reconstruction. Aggressive debridement and irrigation are usually the first line of therapy, followed by systemic antibiotics and placement of occlusive dressings, vacuum-assisted wound closure devices, or temporary prosthetic patches in order to stabilize the patients wound prior to definitive reconstruction.

If the abdominal defect is complex and there is fear of entering the bowel, the patient should undergo preoperative bowel preparation. All patients should receive prophylactic antibiotics within a half hour of operative incision.

Operative Repair

The goals of managing abdominal wall defects are to restore structure and function to the abdominal wall musculature and to create long-lasting wound coverage. Primary repair of abdominal wall hernias can be done when the defect is small, usually defined as $\leq$2 cm in diameter, and there is adequate surrounding tissue. Larger defects measuring >2–3 cm in diameter have a high recurrence rate if closed primarily and thus are better repaired with mesh. Primary repair, even of small hernias with defects <3 cm, is associated with high recurrence rates between 10 % and 50 %. This can be reduced by more than half with the use of prosthetic mesh. Options for mesh placement are onlay patch superficial to the fascia as a buttress for a primary tissue repair, placement within the fascial defect to bridge the gap between the edges, intraparietal within the abdominal wall muscular or aponeurotic layers or as an intraperitoneal underlay patch deep to the fascia.

A variety of synthetic mesh products are available; however, no ideal mesh exists. It must be hypoallergenic, noncarcinogenic, create minimal inflammation, and remain resistant to mechanical stress while still being compliant and dynamic as part of the abdominal wall. A mesh is more susceptible to infection than autologous tissue and has an increased propensity to erode through adjacent structures. Advantages of mesh include wide availability, lack of donor site morbidity, and its strength.

Polypropylene mesh is most commonly used. It is permanent, somewhat flexible, and porous, which allows for ingrowth of host fibroblasts, and thus is easily incorporated into the surrounding fascia. There is a risk of forming an enterocutaneous fistula when placed directly on the bowel, so this is avoided unless omentum is interposed between the mesh and viscera.

Polytetrafluoroethylene, or PTFE, is flexible and smooth. Fibroblast proliferation still occurs through pores, but PTFE does not absorb fluid, and thus it is not incorporated into the host fascia. Because of PTFE's resistance to adherence, it becomes infected more easily than polypropylene, and when infected, PTFE almost always must be removed. Seromas form more easily with the use of PTFE. Vicryl™ mesh is a tightly woven mesh that is thick and flexible with high tensile strength. It is an absorbable mesh that is completely hydrolyzed in 90–120 days. This allows for temporary wound coverage and abdominal wall support in a contaminated field, and thus this type of mesh is usually used in staged abdominal reconstructive procedures. Composite mesh products combine the attractive qualities of nonabsorbable and absorbable mesh or nonabsorbable mesh with mesh that can be placed against the viscera. The most common composite mesh merges polypropylene and PTFE by layering the two on top of one another. The PTFE side serves as a protective interface that is safe to lay against the bowel, while the polypropylene side faces anteriorly to be incorporated into the fascia.

The newest development is a nonsynthetic or natural tissue mesh. These products are composed of acellular collagen harvested from porcine intestinal submucosa or dermis, or they can be made from cadaveric-derived acellular dermal tissue matrix, both of which lack all antigenic cellular elements. These mesh materials can be treated to cross-link collagen, which increases their strength and durability at the expense of fibroblast ingrowth. With time, these meshes are integrated into host tissue, remodeled, and replaced by host collagen. These biosynthetic meshes are more expensive alternatives, and no data exists to suggest they are any more effective than conventional meshes, so they are currently used when there is active infection or significant contamination.

The laparoscopic approach for abdominal wall hernia repair has been increasing but is usually reserved for large or recurrent defects, as it requires general anesthesia, in contrast to the open technique, which can be performed under local anesthesia. Scope and trocar placement are inconsistent and depend on the size and location of the hernia. The hernia contents are reduced, adhesions are lysed, and a piece of mesh is stapled/sutured into place with at least 4 cm of overlap around the defect using an intraperitoneal underlay technique. The advantages of a laparoscopic approach are quicker recovery time and less postoperative pain, as well as the ability to examine the undersurface of the abdominal wall to reveal other defects that might not otherwise be found and repaired. The incidences of postoperative complications, infections, and recurrences are less in hernias repaired laparoscopically.

Massive abdominal wall defects can be particularly difficult. One option for repair is to perform a staged reconstruction using a composite mesh for patients with loss of abdominal domain and lateral retraction of the abdominal wall musculature. The initial stage involves reduction of the hernia and placement of a large sheet of composite mesh that is secured to the fascial edges with a running suture. Subsequent stages involve serial elliptical excisions of the mesh until the fascia can be approximated in the midline without tension. Eventually, the fascia is reapproximated in the midline with an onlay mesh patch if needed. Another option for the repair of complex or large defects is the separation of components technique. Several options exist for this technique, but a common methodology involves separation of the lateral muscular layers of the abdominal wall to allow their advancement toward the midline. By raising large subcutaneous flaps lateral to the fascial defect above the external oblique fascia that carry past the linea semilunaris, some advancement of the abdominal wall can be gained. A relaxing incision is made on the lateral external oblique aponeurosis, and the external oblique is then separated from the internal oblique, allowing its advancement. These flaps, when performed on both sides of the abdominal wall, can produce up to 20 cm of mobilization at the waistline. This technique allows tension-free closure of these large defects; however, recurrence rates of up to 30 % have been documented without the use of a reinforcing mesh. Components separation is associated with a higher risk of wound infection, but it is a useful technique to avoid the use of prosthetic materials and achieve a definitive repair under much less tension than a primary repair.

Complications

Mesh Infection

Mesh infections are serious complications after a hernia repair. If PTFE becomes infected, it must be removed, although this option requires closure under tension leading to likely recurrence of the hernia. In open ventral hernia repair, wound and mesh infections are not infrequent. The laparoscopic technique leads to a low rate of wound complications, and mesh infections occur in less than 1 % of cases.

Seromas

Seroma formation can occur after both laparoscopic and open hernia repair, although there is an increased risk of seroma formation with an open repair. In large open ventral hernia repairs, drains are often placed to obliterate any dead space, but these drains can cause mesh contamination, and seromas can form after drain removal. During a laparoscopic repair, the hernia sac is not resected, so a seroma will frequently result, although most resolve over time. If the seroma persists for more than 6–8 weeks, aspiration can be considered.

Enterotomy

Intestinal injury during adhesiolysis is a concerning complication of a hernia repair. Management of an enterotomy depends on whether the small or large bowel was injured and the amount of spillage into the peritoneum. If an enterotomy occurs, options for repair include aborting the procedure, a primary tissue or biologic tissue repair, or a delayed repair using a prosthetic mesh for several days. However, with gross contamination, a prosthetic mesh is contraindicated.

59
Indications for Renal Transplantation

Deepak Mital and Erica Hammes

Indications

Most patients with chronic kidney disease (CKD) are candidates for renal transplantation (Fig. 59.1a, b). Stage 4 or 5 CKD with a creatinine clearance of 20 ml/min or less is the point at which a patient may be listed and accrue waiting time for a deceased donor kidney. These patients are maintained on either hemodialysis or peritoneal dialysis. Patients with an acceptable living donor may receive a "preemptive" transplant without ever starting dialysis, thus avoiding the risks associated with chronic dialysis (Table 59.1).

Causes of CKD are numerous (Table 59.2), with the overwhelming majority caused by diabetes and hypertension. Focal segmental glomerulosclerosis (FSGS) recurs in 20–50 % of renal transplants, and IgA nephropathy does so in greater than 50 %. In the latter, administration of fish oil (omega-3 fatty acids) posttransplant can decrease recurrence. For patients with a history of cancer, the *Israel Penn International Transplant Tumor* Registry[1] is a good resource for determining when a patient may be eligible to receive a transplant.

Patients with SLE may have high levels of autoantibodies, which can lead to a false-positive crossmatch. The disease usually remains quiescent after a transplant due to immunosuppression. It is important to look for anticardiolipin and other lupus antibodies, which may cause vascular thrombosis due to a hypercoagulable syndrome.

Evaluation, Recipients

The patient undergoes a history and physical (to determine functional status and physiologic problems that need further workup). Advanced age is not a contraindication to a transplant, but extra caution is exercised for patients older than 75 years. Serologic tests, CBC (anemia, leukopenia), chemistry panel (PTH levels, amylase/lipase, blood sugar), and liver enzymes (hepatitis) identify conditions needing more detailed evaluation and management (Table 59.3). A positive result is not always a contraindication.

Living Donors

Living donors are evaluated to identify possible disease states that may be transmitted to the recipient. The donor must be at least 18 years old and free of coercion. Donors may be related (living related transplant; LRT) or unrelated/altruistic. They must be free of hypertension and diabetes, although there are exceptions at certain centers for older hypertensive donors well controlled on a single medication. All donors must undergo a complete psychosocial assessment and be evaluated by an independent nephrologist not involved in the patient's care.

Donors undergo a magnetic resonance angiogram (MRA) or computed tomography (CT) scan of the abdomen to determine renal anatomy. It is best to use kidneys with single vessels, preferably the left kidney, which has a longer renal vein.

Kidney Paired Donation

Kidney paired donation (KPD) is increasingly common for altruistic and related donors who are ABO-mismatched or have a positive crossmatch. This may result in a donor exchange with a chain of multiple transplants, often across different centers and states.

Cadaveric Donors

Cadaveric donors undergo a similar assessment to determine kidney function. They must have confirmed brain death, which is usually a result of trauma or cerebrovascular

[1] www.ipittr.org

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_59, © Springer New York 2015

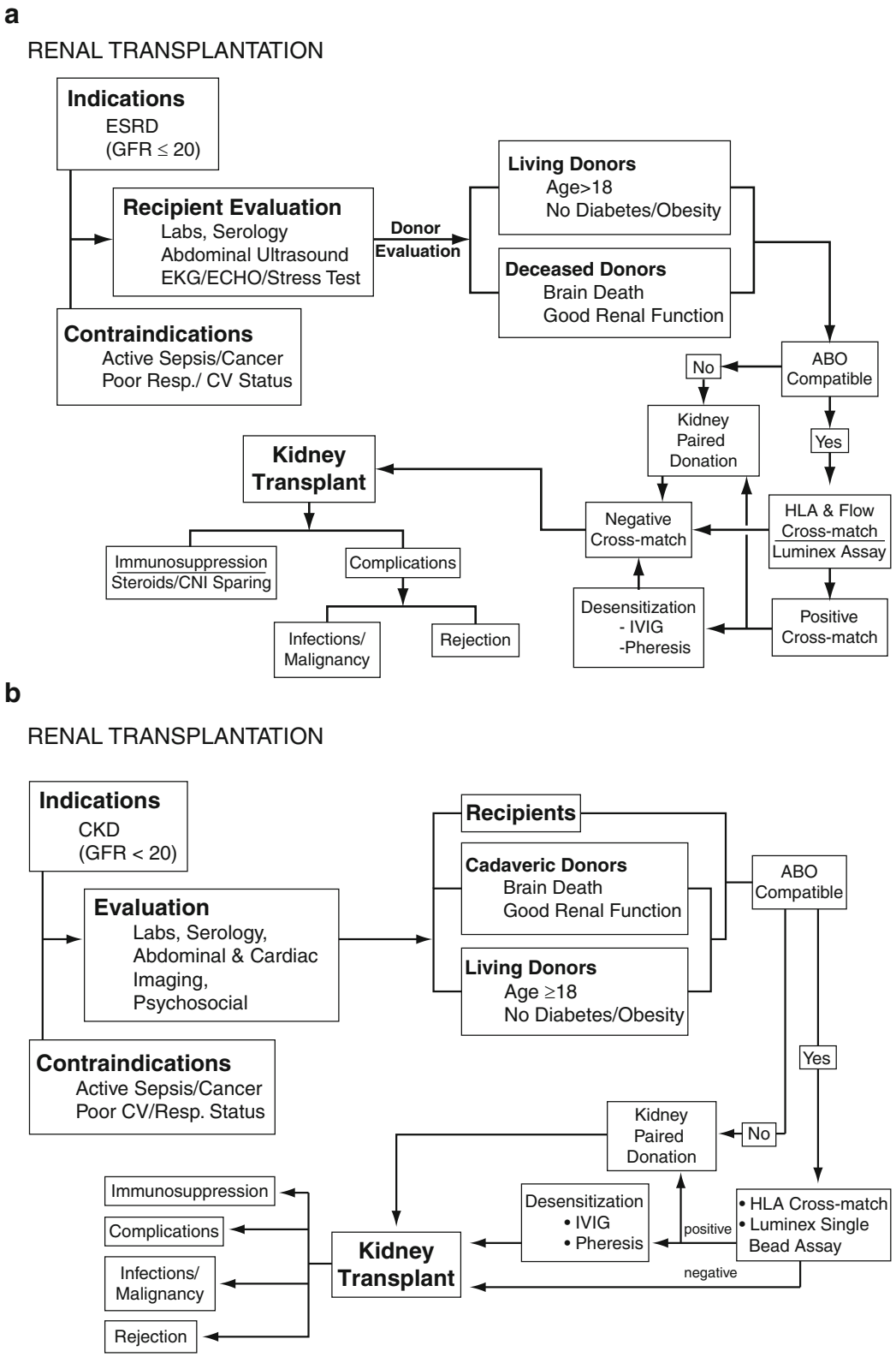

FIG. 59.1 Algorithm for renal transplantation. (**a**) End-stage renal disease (*ESRD*). (**b**) Chronic kidney disease (*CKD*). *FSGS* focal segmental glomerulosclerosis, *GFR* glomerular filtration rate, *EKG* electrocardiogram, *CV* cardiovascular, *HLA* human leukocyte antigen, *IVIG* intravenous immunoglobulin, *CNI* calcineurin inhibitor, *KPD* kidney paired donation

TABLE 59.1. Long-term complications of CKD.

Left ventricular hypertrophy	Coronary artery disease
Increased mortality rate	Vascular calcification
Anemia	Renal osteodystrophy
Peripheral neuropathy	Increased risk of MI
Increased risk of cancer (renal, urinary tract)	

TABLE 59.2. Etiology of CKD.

Metabolic	**Primary glomerulonephritis**	**Hereditary**
Diabetes	FSGS	Polycystic kidney disease
Vascular disease	Membranous nephropathy	Hereditary nephritis (Alport's)
Hypertension	IgA nephropathy	Tuberous sclerosis
Renal artery stenosis	IgM nephropathy	Fabry's disease
Renal vein thrombosis	Postinfectious GN	1° Oxalosis
Malignancy	**Secondary glomerulonephritis**	Renal hypoplasia/ dysplasia
Renal tumors	Systemic lupus erythematosus	Prune belly
Multiple myeloma	Henoch-Schonlein syndrome	Drash syndrome
Amyloidosis	Hemolytic uremic syndrome	
Miscellaneous	Scleroderma	
Sickle cell disease	Wegener's granulomatosis	
Postpartum renal failure	Good Pasteur's syndrome	
AIDS nephropathy	Substance abuse nephropathy	

TABLE 59.3. Pretransplant testing and management.

Positive serologic test	Management pretransplant
CMV	IV ganciclovir, oral valganciclovir
EBV	IV ganciclovir, oral acyclovir
VZV	Acyclovir, varicella immune globulin, varicella vaccine
Hepatitis C	Interferon, ribavirin, many new options
HIV	HAART, monitoring CD4 counts
TB	Isoniazid, rifampin

accident (CVA). The donor is maintained on a ventilator and the heart is usually beating until the organs are procured. Donors must lack hypotension (damage to the kidneys), be free of high-dose pressor agents, lack elevated serum creatinine, oligoanuria, and trauma to other organs. There is an increase in utilization of DCD (donation after cardiac death) and ECD (expanded criteria donors: aged >50 with elevated serum creatinine, history of hypertension, or death due to CVA) donors due to national organ shortage. Current waiting times are more than 6 years in many states.

The kidneys are flushed with Viaspan (University of Wisconsin) solution, which resembles intracellular fluid. If the "cold ischemia time" (CIT) prior to transplantation exceeds 24 h, the incidence of acute tubular necrosis (ATN) rises exponentially. With pulsatile-perfusion machine preservation, ATN rates are significantly lower with CIT > 24 h.

Contraindications

1. *Sepsis*.
2. *Advanced cardiovascular disease*. Left ventricular ejection fraction <30 %, severe bilateral iliac/femoral artery atherosclerosis, and pulmonary hypertension may be contraindications for transplant.
3. *Poor respiratory status*, e.g., needing O_2 therapy at rest.
4. *Cancer*. Completely excised skin cancers are exceptions.
5. *AIDS*. Some patients who have no detectable viremia on antiretroviral therapy are now being considered candidates. CD4 counts are carefully monitored.
6. *Psychosocial*. Active drug abuse, psychosis, and a lack of social support systems predict noncompliance with medications and follow-up care.

HLA Matching and Crossmatch

ABO blood group compatibility is essential before considering human leukocyte antigen (HLA) matching. Otherwise, the kidney will be destroyed by the preformed antibodies within hours.

Human leukocyte antigens are located on the short arm of chromosome 6 within the major histocompatibility complex (MHC) and indicate genotypic matching. *A, B, and DR are the loci considered, with more weight being given to the DR mismatching*. A child inherits one haplotype from each parent; thus, parents are considered a one-haplotype match. A patient's siblings may be HLA identical (25 % chance), share one haplotype (50 % chance), or have no HLA matching (25 % chance). Long-term results are the best for HLA-identical transplants.

Luminex bead single-antigen analysis and flow cytometric crossmatch (FCXM) are more sensitive for detecting HLA antibodies than the previously used complement-dependent cytotoxicity (CDC) assay. If the crossmatch is "positive" (indicating antibody-mediated rejection of donor cells), another donor must be found or, more recently, techniques to desensitize these donors prior to transplantation must be employed. Immunomodulation using intravenous immunoglobulin (IVIG) and plasmapheresis may allow transplant despite a positive crossmatch. The data from crossmatches across the country is pooled in a collective database for deceased donor organ allocation by the United Network for Organ Sharing (UNOS) .[2]

[2] www.unos.org

Transplant

In the perioperative period, a chest X-ray and electrocardiogram are obtained along with a medical and laboratory assessment. Prophylactic antibiotics and immunosuppressants are administered. The kidney is transplanted retroperitoneally in the iliac fossa. The renal artery and vein are anastomosed to the external iliac vessels in an end-to-side fashion. The ureter is anastomosed to the bladder, usually with a stent in place. The native kidneys are not removed unless there is a history of recurrent pyelonephritis. The usual hospital stay is 4–5 days.

Immunosuppression

Induction immunosuppression (perioperatively) may include monoclonal or polyclonal antibodies. Maintenance immunosuppression (posttransplant) is with a combination of calcineurin inhibitors (CNIs) (e.g., tacrolimus/cyclosporine) and antiproliferative drugs such as mycophenolate mofetil (CellCept) or sodium (Myfortic). Corticosteroids are being avoided or withdrawn early due to their well-known side effects of insulin resistance, weight gain, and osteoporosis. Other classes of drugs include mTOR inhibitors such as sirolimus. Newer biologic agents such as anti CD25 and CD40 monoclonal antibodies are also being developed.

Complications

Surgical complications include vascular thrombosis/stenosis (<2 %). This is usually due to technical error or a hypercoagulable syndrome. A thrombosed graft is often lost due to cortical necrosis resulting from the ischemia.

Ureteric leaks or stenoses (1–5 %) are usually due to technical error/ischemia. They may be treated by placing a stent (interventional radiology) or surgical correction. Lymphoceles (5–15 %) are usually caused by leakage of lymph from the lymphatics around the iliac vessels that were not ligated at the time of transplant. The fluid collection may compress the ureter leading to obstruction. Treatment consists of percutaneous drainage or surgical marsupialization into the peritoneum, often done laparoscopically.

Infections and Malignancies

Infections and malignancies may occur after transplantation. Bacterial infections (central lines, wound, and urinary infection) are due to perioperative factors and occur in the first 4 weeks posttransplant. Viral infections (>1 month: cytomegalovirus, Epstein-Barr virus, and hepatitis) reflect over-immunosuppression along with transmission of viruses from the donor. Hence, antiviral prophylaxis is given for 3–6 months posttransplant. Opportunistic infections (late, >3 months, e.g., pneumocystis pneumonia/fungal/legionella) are due to prolonged over-immunosuppression, either due to prolonged antibody "induction" therapy after transplantation or to treatment of rejection episodes along with corticosteroids. Posttransplant lymphoproliferative disorder (PTLD) is an over-proliferation of B cells and may be due to EBV infection or over-immunosuppression. Decreasing immunosuppression may help, and PTLD may also be treated with monoclonal anti-B cell agents. Other malignancies are also more common in transplant recipients after years of immunosuppression.

Rejection

Rejection may follow one of three patterns:

1. *Hyperacute*: Leads to immediate graft loss within 6 h. It is due to preformed anti-ABO or HLA antibodies. It is rare as it can be predicted by a pretransplant crossmatch.
2. *Acute*: Occurs in 5–20 % of renal transplants. It is most common in the first 6 months to 1 year posttransplant but may occur years later if the patient omits taking antirejection medication. It is T cell-mediated and may be treated successfully in >85 % of cases with a 10–15 mg/kg bolus of IV Solu-Medrol (methylprednisolone) given daily for 3 days. If this fails, anti-T cell antibodies are usually successful.
3. *Chronic*: Interstitial fibrosis-tubular atrophy (IF-TA) is a condition caused by repeated acute rejection episodes, CNI nephrotoxicity, donor injury at time of procurement/transplant, or preexisting donor diseases. There is no successful treatment. Eliminating or lowering the CNI dose and adding sirolimus or CellCept are new strategies that may help.

Results

One-year allograft survival is now >95 % for living donor renal transplant and ~90 % for cadaveric transplants. Five-year allograft survival is best with HLA-identical transplants (~80 %) and worst for poorly matched cadaveric transplants with acute tubular necrosis (<50 %). Results from all US centers are reported in the Scientific Registry of Transplant Recipients.[3] A successful transplant affords these patients a longer and healthier life than on dialysis.

[3] www.srtr.org

60
Indications for Liver Transplantation and Pretransplant Evaluation

Cynthia L. Leaphart and Dana K. Perry

Introduction

Liver transplantation began with experimental canine transplants in the late 1950s. In 1963, the first human liver transplant was attempted by Thomas Starzl. During the first decade of liver transplantation, survival rates were low. The introduction of cyclosporine for immunosuppression, advances in surgical technique, progress in anesthesia and critical care management, and advances in preservation solutions have rapidly accelerated progress. Liver transplant is the treatment of choice for a select group of patients with acute and chronic liver diseases (Fig. 60.1).

In 2011, 16,876 patients were listed for liver transplant but only 5,840 donors were available. Since demand for donor organs far exceeds supply, careful pretransplant evaluation is critical to ensure that a candidate is appropriately selected by disease type, chance of recovery, and adequate psychosocial support after transplantation.

Listing for Transplantation

The United Network for Organ Sharing (UNOS), established in 1986, is a private nonprofit organization under federal contract that was devised to address the nation's critical organ shortage. UNOS has established a system to collect, store, and match patients waiting for organs. UNOS monitors allocation and outcomes while enforcing listing parameters to maximize number of organs transplanted.

To qualify for listing, a patient's expected survival should be 90 % or less within 1 year if a transplant is not performed. Patients listed for liver transplant are ranked according to their MELD (model for end-stage liver disease) score. The MELD score is a calculation based on the patient's bilirubin, INR, and creatinine. Hepatocellular carcinoma (HCC), cholangiocarcinoma (CC), hepatopulmonary syndrome (HPS), familial amyloidosis, and primary oxaluria are case exceptions that receive additional points outside of the MELD scoring system. These exception points are granted on the basis that the underlying liver disease is highly likely to cause death, but the severity of the disease is not captured by the MELD calculation.

Indications for Transplantation

The liver is susceptible to damage from metabolic, toxic, circulatory, microbial, and neoplastic causes. The clinical impact of early liver disease is often masked by the functional reserve of the liver. As scarring progresses, hepatocyte destruction and parenchymal damage lead to hepatic failure and decompensation, such as gastrointestinal bleeding, electrolyte disturbances, coagulopathy, ascites, malnutrition, encephalopathy, and infection. In some patients, liver transplant is the only treatment option.

End-stage liver disease is categorized as acute or chronic. **Acute liver failure**, or fulminant hepatic failure (FHF), is defined as the development of hepatic encephalopathy and coagulopathy within 8 weeks of symptom onset in patients without a preexisting diagnosis of liver disease. *The most common causes of acute liver failure are acetaminophen toxicity, acute hepatitis B infection, medications and chemicals, Wilson's disease, and microvesicular steatosis syndromes (e.g., acute fatty liver of pregnancy)*. The mortality rate is high without transplant. Patients presenting FHF require more intensive care pretransplant due to the higher incidence of cerebral edema causing neurologic complications, coagulopathy, bacterial and fungal infections, and renal failure. *Chronic causes of liver disease that are amenable to treatment by liver transplant may originate from viral infections, metabolic errors of metabolism, drug or toxin exposure, cholestasis, circulatory disorders, autoimmune problems, or neoplasia*.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_60,

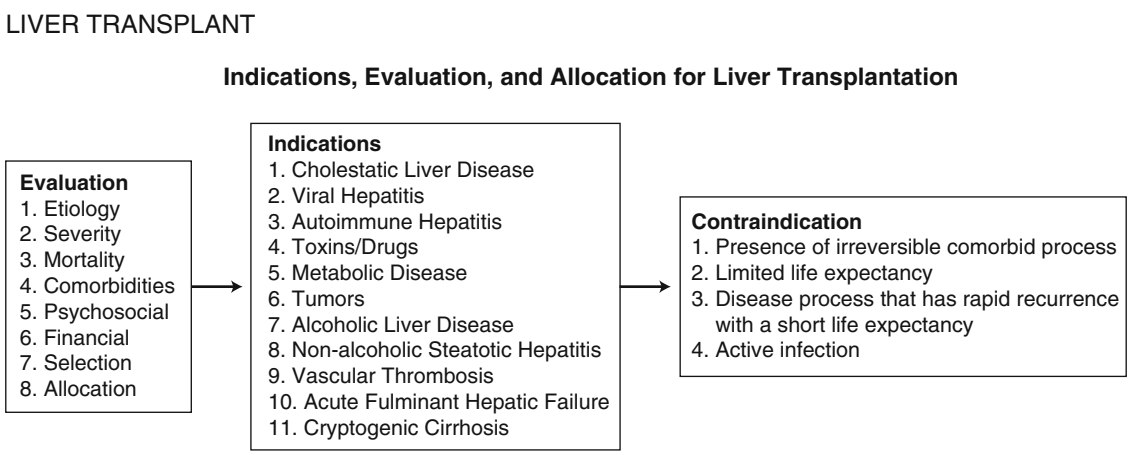

FIG. 60.1 Indications, evaluation, and allocation for liver transplantation

Viral Hepatitis: Hepatitis B or Hepatitis C

Hepatitis C is the leading cause of end-stage liver disease requiring transplant in the United States. In the pretransplant period, treatment of the disease and reduction of the viral load with antiviral strategies are attempted if feasible. Patient survival after transplant is negatively affected by pretransplant viral load, cytomegalovirus status, advanced donor and recipient age, hyperbilirubinemia, and elevated INR. Unlike hepatitis C, hepatitis B is not frequently associated with graft reinfection due to the availability of hepatitis B immune globulin prophylaxis and use of tolerable oral drug therapy.

Alcoholic Liver Disease

Chronic alcohol consumption adversely affects the liver by lipid deposition, which progresses to fibrosis, hepatocyte damage, and inflammatory reaction, causing irreversible progression to a cirrhotic state. Risk factors include daily consumption of alcohol, consuming multiple drinks at a single sitting, drinking outside meals, genetic factors, obesity, and advancing age. Patients are evaluated by Psychiatry and Addiction Medicine physicians and should demonstrate a period of abstinence. A period of abstinence is required because liver disease can improve deeming transplant unnecessary, it can identify those at risk for recidivism, and allow time for addiction counseling.

Cholestatic Diseases

Primary biliary cirrhosis (PBC) is a nonsuppurative, granulomatous destruction of medium-sized intrahepatic bile ducts leading to portal inflammation and cirrhosis. Primary sclerosing cholangitis (PSC) is inflammation, obliterative fibrosis, and segmental constriction of both intrahepatic and extrahepatic bile ducts. Both PBC and PSC patients undergo frequent endoscopic and percutaneous procedures and are susceptible to resistant bacterial and fungal infections that may accelerate mortality.

Malignant Tumors

Primary liver cancers arise from hepatocytes (HCC) or bile duct epithelium (CC) and can be cured with liver transplantation in select patients. Posttransplant 4-year patient survival is 85 % for patients chosen for transplant with HCC according to the Milan criteria (one tumor<5 cm or one to three tumors, each<3 cm, absence of vascular or lymphatic invasion, no extrahepatic disease). Transplantation is indicated for early stage unresectable hilar CC after neoadjuvant radiotherapy and chemotherapy. Patients with CC must undergo pretransplant laparotomy to sample hilar lymph nodes assuring no metastatic disease. Patients that receive MELD exception points must fit in the respective criteria.

Nonalcoholic Steatotic Hepatitis (NASH)

NASH is a major cause of liver failure and is predicted to be the primary cause of cirrhosis and liver transplant in the United States in the future. Cirrhosis is due to fatty infiltration and subsequent inflammation of the liver. Careful evaluation is warranted in NASH patients who often have metabolic syndrome. Mortality tends to be higher in the early postoperative period, although overall outcome is similar to other patients undergoing transplant.

Metabolic Diseases

Hereditary hemochromatosis, alpha$_1$-antitrypsin deficiency, amyloidosis, and Wilson's disease are inborn errors of metabolism that result in chronic liver injury and cirrhosis.

Vascular Abnormalities

Budd-Chiari syndrome is an acute and fatal thrombotic occlusion of the hepatic veins characterized by hepatomegaly, weight gain, ascites, and abdominal pain.

Liver Transplant Evaluation

Patients in the aforementioned categories are referred to transplant centers for liver transplantation evaluation. The evaluations are performed by a team of hepatologists, surgeons, psychiatrists, infectious disease physicians, social service workers, and finance specialists. Cardiologists and pulmonologists may also be involved. The complex evaluation selects patients that can survive the perioperative period, comply with an advanced medical regimen, and have good psychosocial support.

Transplant evaluation begins with basic laboratory assessment of chemistry, hematology, viral serologies, blood type, and baseline evaluation of alpha-fetoprotein, carcinogenic embryonic antigen, or CA 19-9, if malignancy is present. Neurologic evaluations are obtained in patients with a history of stroke or cognitive dysfunction. Pulmonary evaluation includes room air arterial blood gas measurements, chest X-ray, and pulmonary function tests. Cardiac evaluation includes an electrocardiograph, echocardiogram with a bubble study, and cardiac stress test if greater than 50 years old or with other coronary risk factors. Angiography or right heart catheterization is ordered for positive stress tests or elevated pulmonary artery pressures, respectively. Upper and lower endoscopy is performed to assess signs of portal hypertension and colorectal cancer, respectively. Abdominal imaging assesses the liver, its vasculature, and potential malignancies with ultrasound, CT, and/or MRI. Patients with HCC or CC will undergo CT scans of the chest and bone scans to access for metastatic disease. In females, a pap smear is required within 1 year of listing, and mammography is performed for patients older than 40 years of age. A PSA and prostate exam is performed in men.

Psychosocial assessment is critical in transplant selection and management. Economic barriers must be evaluated and overcome to prevent graft failure from lack of follow-up and poor medication compliance. Adequate resources should be in place to enable success after liver transplantation.

Part VI
Surgical Oncology

61
Palpable Breast Mass

Rosalinda Alvarado

Introduction

Palpable breast masses are very common and tend to be benign. However, it is important to thoroughly examine the patient and decide what diagnostic testing is necessary to rule out malignancy. The likelihood of a mass being malignant depends on the patient's age, history, risk factors, appearance on examination, and imaging characteristics. The advantages and limitations of imaging modalities (mammograms, ultrasound, magnetic resonance imaging [MRI]) must be appreciated before implementing them into practice. Mammography, for example, is very useful in women over 40 years but can be difficult to interpret in younger women secondary to the density of their breasts. Conversely, MRI is highly sensitive in the detection of lesions in the breast and as such can lead to a high false-positive rate, thereby subjecting women to potentially unnecessary surgery.

History

The history should include specific questions pertaining to the mass, including length of time it has been present, association with the menstrual cycle, tenderness, and growth over time. Risk factors for breast cancer include, but are not limited to, long period of uninterrupted menstrual cycles, nulliparity, use of hormone replacement therapy, history of radiation, family history, personal history of breast cancer, and previous breast biopsies showing atypical hyperplasia or lobular carcinoma in situ. Age certainly needs to be taken into account as breast masses are more likely to be malignant as the patient ages (Fig. 61.1a–c). A strong family history might compel a clinician to request more diagnostic testing in a young woman who otherwise would have been felt to have a benign fibroadenoma. It is also important to know when genetic testing is indicated as this will guide future surveillance and management.

Physical Exam

A comprehensive breast examination should include both breasts, the chest wall, axilla, and cervical, supraclavicular, and infraclavicular nodal basins. Ideally this should be done 3–10 days after the onset of menses. The examination should begin with the patient upright in order to visually inspect for asymmetry, skin changes, obvious masses, and/or nipple inversion/retraction. While upright, the breasts and nodal basins should be palpated. Then the patient is placed supine with one arm raised and the clinician thoroughly palpates each breast and the nodal basins. The nipple-areola complex should be examined carefully for skin changes and discharge. It is important that the examination be done in both positions as some masses may become more pronounced in one position versus the other. The size of a mass should be documented.

Diagnostic Evaluation

Mammography should be considered first in all patients; however, interpretation may be difficult in very young women (under 30 years). Digital mammography has proven to be more useful in younger women than standard mammography, and it is useful as a baseline study. Ultrasound is a very vital adjunct to mammography and physical examination; it can quickly differentiate between a solid and cystic mass and it may visualize masses that were not evident on mammogram. Ultrasound can further characterize the mass as malignant or benign appearing using the Breast Imaging-Reporting and Data System (BI-RADS) classification. MRI is also a helpful tool, particularly when a palpable breast mass is not appreciated on mammogram and ultrasound. With the data thus obtained, the clinician can decide whether a biopsy is warranted or not.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_61, © Springer New York 2015

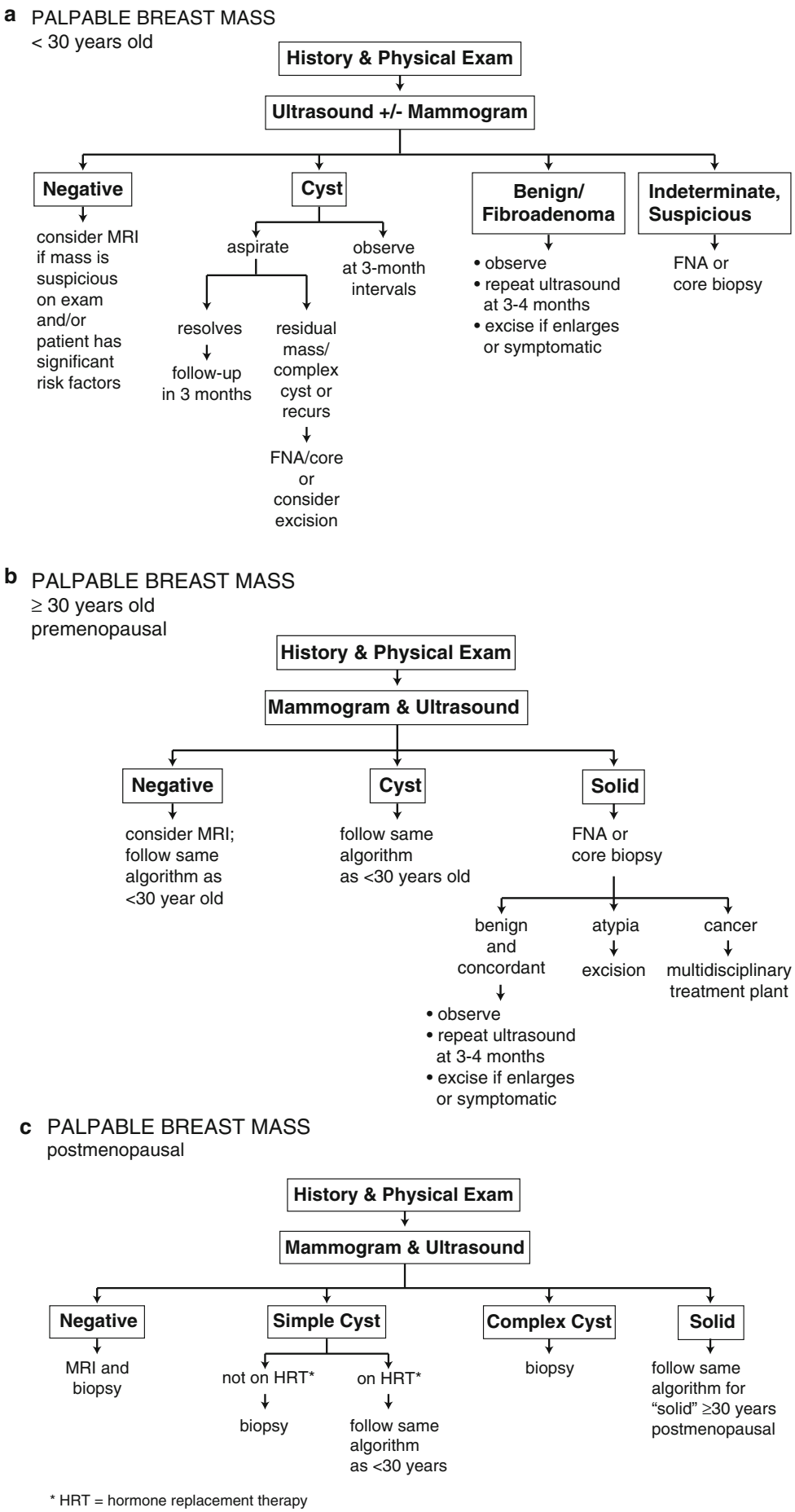

FIG. 61.1 Algorithm for palpable breast mass. (**a**) Patient younger than 30 years. (**b**) Patient older than 30 years and premenopausal. (**c**) Patient postmenopausal. *MRI* magnetic resonance imaging, *FNA* fine needle aspiration, *HRT* hormone replacement therapy

Women Under 30 Years

The mass is most likely of benign etiology such as a fibroadenoma or cyst (Fig. 61.1a). Special attention should be given to a personal history of breast atypia or a family history of breast cancer in this age group, as this will help guide how aggressive one should be with diagnostic testing. A young woman with a BRCA mutation should be addressed differently than one with no family history or genetic mutation. Although a mammogram may be considered, an ultrasound will be most useful. A cyst will have a characteristic appearance (anechoic, well circumscribed, and compressible). A fibroadenoma will appear homogenous, hypoechoic, and well circumscribed. If the ultrasound is consistent with benign pathology, a follow-up clinical examination and ultrasound should be done in 3–4 months (sooner if the mass becomes symptomatic or rapidly enlarges). Alternatively, a large or symptomatic cyst can be aspirated under ultrasound guidance. *A biopsy should be considered in cases where imaging does not clearly characterize the mass*. A core needle biopsy is a good method as it provides tissue architecture and can confirm benign pathology or diagnose malignancy, differentiating between in situ and invasive cancer. Hormone receptor status can also be determined on core needle biopsy.

Premenopausal and Over 30 Years

A breast mass in this age group is usually a cyst (Fig. 61.1b). However, it is important to note that the chance of breast cancer is higher here than in the previous age group. Mammography and ultrasound should both be considered. If it is clearly cystic or if there are multiple cysts, the patient can be followed with clinical breast exams and imaging. If the cyst is large or symptomatic, it should be aspirated. It is not necessary to send the fluid for cytologic studies unless it is a recurrent or complex cyst. If a cyst recurs after aspiration, or it is complex, excision should be considered. Solid masses in this age group that clearly appear benign can be followed both clinically and with imaging in 3–4 months. When the solid mass is suspicious or clearly malignant appearing, a core needle biopsy should be obtained, under image guidance if necessary. If benign tissue is obtained *and this is concordant with imaging*, the patient can be followed clinically and with imaging. Alternatively, the patient may desire excision if the mass is symptomatic or enlarging. Pathology consistent with malignancy should be followed by appropriate staging and a multidisciplinary treatment plan.

Postmenopausal

The incidence of breast cancer in this age group is highest (Fig. 61.1c). As such mammography and ultrasound should be used in all of these patients with new palpable breast masses. A biopsy should be considered in all patients with palpable breast masses in this age group. If biopsy results are inconclusive or discordant with imaging studies, biopsy should be attempted again by an experienced radiologist or surgeon, or strong consideration should be given to an excisional biopsy. Every attempt should be made at obtaining a diagnosis prior to reaching the operating room.

In general, breast biopsy should always be considered when the breast mass appears suspicious/malignant on imaging, appears in a patient with numerous risk factors for breast cancer, is enlarging clinically or on imaging, or when there is a concern about patient compliance or follow-up. Any patient who does not undergo excision of a mass should be followed both clinically and radiographically.

62
Abnormal Mammogram

Katherine Kopkash

Screening

Screening patients for breast cancer with mammography should start at age 40 and continue annually as long as the patient is in good health. For women at high risk for breast cancer, most medical societies recommend annual mammography as well as a magnetic resonance imaging (MRI) scan starting at age 35.

A screening mammogram usually includes two views, a craniocaudal and a mediolateral oblique view. If screening mammogram reveals an abnormality or if there is a suspicious breast-related symptom or sign (e.g., mass, skin change), a diagnostic mammogram is performed (Fig. 62.1). This involves the standard views plus additional angles and special views that often include magnification to enlarge the concerning areas of the breast. Diagnostic mammography is also performed on patients with implants and patients who have been treated for breast cancer. Mammographic findings concerning for cancer can be divided into two categories, major and minor. **Major** findings concerning for malignancy include a mass with spiculated margins and/or the finding of clustered microcalcifications. **Minor** signs concerning for malignancy include a poorly defined mass, microlobulations, architectural distortion, asymmetric density, nipple retraction, and axillary adenopathy. After the radiologist interprets the imaging, he or she will assign a Breast Imaging-Reporting and Data System (BI-RADS) score. These scores standardize the reporting of mammographic findings as well as offer recommendations for further work-up (Table 62.1).

Other Diagnostic Modalities

Breast ultrasound is not well established as a screening modality but serves a significant role as a diagnostic modality. It can delineate the shape, orientation, margin, boundary, echogenicity, and posterior acoustic characteristics of a mass. Ultrasound is used to determine if palpable masses are cystic or solid. When cysts are encountered, ultrasound characteristics can help determine if they are simple or complex and therefore if a biopsy is needed. Abscesses of the breast also have specific characteristics seen on ultrasound, and their drainage can be facilitated by ultrasound guidance. Ultrasound guidance is especially helpful when performing needle or core biopsies of lesions. Image-guided biopsy of a lesion is superior to open excisional biopsy in terms of accuracy, lower cost, less pain, and better planning for the definitive surgery.

Breast MRI has been used with increasing frequency over the past 20 years and is now the screening modality of choice for BRCA-positive patients. It is commonly used to study indeterminate lesions found on mammogram and ultrasound. MRI is also used to evaluate patients presenting with positive axillary lymph nodes but disease that is otherwise occult on mammography and to evaluate response to neoadjuvant chemotherapy.

Biopsy

Tissue biopsy is recommended for any patient with a BI-RADS 4 or 5 categorization on mammography. If the lesion is palpable, a core biopsy can be performed in the office with local anesthesia under ultrasound guidance if needed. If the lesion is non-palpable, stereotactic core biopsy should be performed. If the lesion was only identifiable on ultrasound or MRI, that modality should be used to perform the biopsy. Core biopsy is preferable to fine needle aspiration as it provides more tissue for assessment. If a patient has a mass or skin changes and normal imaging, the surgeon must consider patient-specific risk factors to determine if a biopsy is warranted (Table 62.2). Ruling out cancer is of the utmost importance, and therefore biopsy is performed in the majority of cases. Lesions that appear on ultrasound as simple cysts or fibroadenomas can be watched safely, but other findings require tissue to determine if they are truly benign. If core biopsy results are discordant with imaging (benign results but a BI-RADS 5 categorization), the physician needs

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_62, © Springer New York 2015

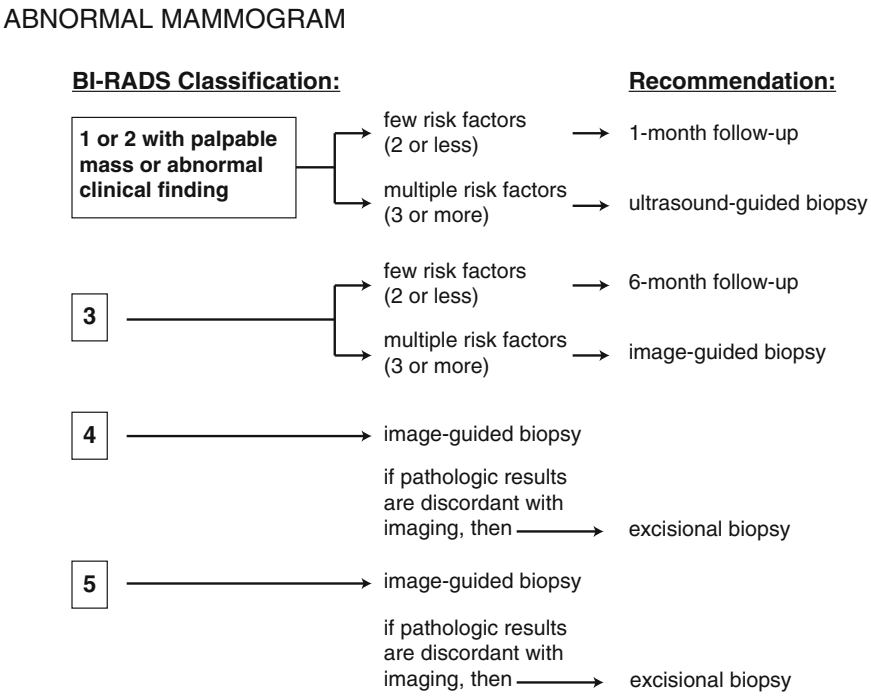

FIG. 62.1 Algorithm for abnormal mammogram. *BI-RADS* Breast Imaging-Reporting and Data System

TABLE 62.1. BI-RADS categorization.

BI-RADS category	Impression	Recommendation
0	Unable to assess	Needs additional imaging
1	Negative	Routine screening
2	Benign finding	Routine screening
3	Probably benign finding	Short term follow-up (usually 6 months)
4	Suspicious abnormality	Biopsy should be considered
5	Highly suggestive of malignancy	Biopsy strongly recommended

to be concerned for sampling error, and an excisional biopsy is recommended. Palpable axillary lymphadenopathy may also be biopsied under ultrasound guidance in the clinic.

Definitive Surgery

One of the goals of breast surgery is for patients to have only one trip to the operating room; biopsy is therefore preferably performed in the clinic or radiology suite. This establishes the diagnosis of cancer or benign disease ahead of time and allows a multidisciplinary team to establish a definitive treatment plan. If the lesion is benign but the patient and her physician feel it should be removed, excisional biopsy can be performed. If cancer is diagnosed, lumpectomy or mastectomy can be performed in combination with a sentinel lymph node biopsy or axillary dissection as appropriate.

TABLE 62.2 Risk factors for breast cancer

Breast cancer risk factor	Specifics
Gender	
Age	
Genetic factors	BRCA carrier, Li-Fraumeni or Cowden syndrome, ataxia telangiectasia
Family history of breast cancer	
Personal history of breast cancer	
Previous breast biopsy	Proliferative breast disease without atypia, atypical hyperplasia, lobular carcinoma in situ
Previous thoracic radiation	
Endocrine risk factors	Early menarche, late menopause, late parity, nulliparity, long-term estrogen/progesterone hormone replacement
Lifestyle factors	Alcohol, obesity

63
Invasive Breast Cancer

Alicia Growney and Katherine Kopkash

Introduction

The estimated number of new cases of invasive breast cancer in 2012 was estimated to exceed 226,000; nearly 40,000 dying from the disease. The vast majority of patients are women with just under 1 % of breast cancer cases affecting men. Factors associated with increased risk include gender, increasing age, early onset menarche, late menopause, nulliparity, late first pregnancy, obesity after menopause, alcohol use, and personal or family history of breast cancer; the most significant among these are female gender and advancing age. The median age at diagnosis is 61.

The lifetime risk of a woman developing breast cancer is approximately 12 %, and this risk doubles if she has a first-degree relative (mother, sister, daughter) diagnosed with the disease.

The genetic predisposition to developing breast cancer through the BRCA 1 or BRCA2 mutations accounts for approximately 5–10 % of cases of breast cancer in the United States. Inheritance of either of these genes confers up to an 80 % lifetime risk of developing a breast cancer. This gene is also linked to increased risk of ovarian cancer.

The most common pathologic subtypes of breast cancer are invasive ductal (80 %), invasive lobular (10–15 %), and mixed ductal/lobular. The overall survival and outcome is the same for these groups stage for stage. Rare histologic subtypes include medullary, mucinous, tubular, adenoid cystic, and metaplastic carcinomas.

After increasing for more than two decades, the incidence of breast cancer continues to be stable since 2004. It is believed this is due to the results of the Women's Health Initiative Study published in 2002, which linked the use of hormone replacement therapy to an increased risk of breast cancer. Breast cancer is the second leading cause of cancer deaths in women with death rates continuing to decrease especially in women <50 years old. The decrease is felt to reflect treatment advances, earlier detection with screening, and increased awareness (Fig. 63.1).

The surgical treatment of breast cancer continues to evolve from the aggressive Halstedian radical mastectomy routinely performed into the 1970s to less invasive breast conservation therapy, which proved to have equivalent efficacy in the NSABP trials lead by Dr. Bernard Fisher. The ACOSOG (American College of Surgeons Oncology Group) Z-0011 trial published in 2010 demonstrates that we can avoid axillary lymph node dissection in certain lymph node-positive patients undergoing breast conservation without compromising outcome.

Stages I, II, and III Invasive Breast Cancer

The role of neoadjuvant chemotherapy is continuing to evolve and is often used prior to surgery for patients with triple negative disease, HER2+ disease, or those with a clinically positive axilla.

Surgical options include breast conservation followed by radiation versus mastectomy. With either surgery, a sentinel lymph node biopsy is performed to assess lymph node status. In patients undergoing mastectomy, reconstructive surgery can be offered that includes tissue expander/implants or tissue transfer (transverse rectus abdominis myocutaneous flap [TRAM flap], latissimus dorsi flap, deep inferior epigastric perforators flap [DIEP flap], gluteal free flap). Often the type of reconstruction depends on the patient's body habitus.

In the 1990s, the concept of the sentinel lymph node biopsy was introduced and has become the standard of practice in staging the axilla in *clinically node-negative* patients without compromising local recurrence or survival rates. The traditional management of a positive sentinel lymph node has been a level I and II axillary lymph node dissection (ALND) for any patient demonstrating one or more involved lymph nodes with metastatic disease, and this is still the recommended practice in women undergoing mastectomy. This management strategy is changing in sentinel lymph node positive women undergoing breast conservation surgery.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_63, © Springer New York 2015

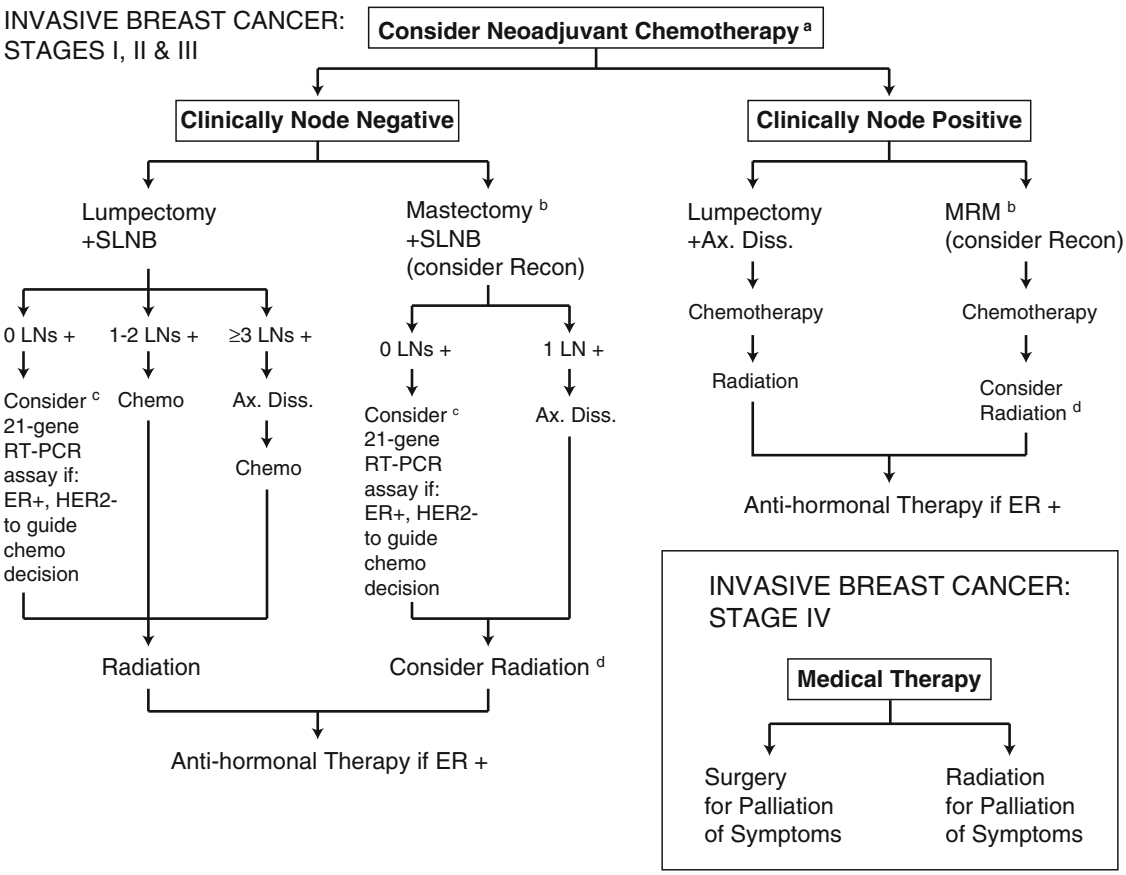

FIG. 63.1 Invasive breast cancer treatment algorithm. *SLNB* sentinel lymph node biopsy, *Recon* reconstruction, *Ax. Diss.* axillary dissection, *MRM* modified radical mastectomy, *LN* lymph node, *ER* estrogen receptor, *RT-PCR* reverse transcriptase-polymerase chain reaction, *Chemo* chemotherapy. [a] = Triple negative cancers, HER 2+ cancers, and patients with a clinically positive axilla. [b] = Indicated for a large tumor-to-breast ratio, multicentric disease, and gene mutation carriers. [c] = Triple negative cancers and HER 2+ cancers will likely receive chemotherapy regardless so assay only indicated for ER+ HER2 patients. [d] = Postmastectomy radiation indicated for large tumors, multiple positive lymph nodes, or tumors invading skin or chest wall

The ACOSOG Z-0011 trial randomized patients with clinically node-negative but hematoxylin and eosin (H&E)-detected positive metastasis in 1 or 2 sentinel lymph nodes to be managed with axillary lymph node dissection or no further directed axillary treatment. All patients went on to receive whole-breast irradiation with a median follow-up of 6.2 years. This study demonstrated no difference in local-regional recurrence rates and no difference in 5-year overall survival or disease-free survival rates. Although the study closed early because of low accrual/event rate, it demonstrated no trend toward clinical benefit of ALND in patients with limited lymph node involvement. A recent survey of the American Society of Breast Surgeons indicates that surgeons are modifying their management of the axilla in patients undergoing breast conservation surgery and performing fewer ALNDs. The morbidity of an ALND (lymphedema, pain, decreased shoulder range of motion, and numbness) can be avoided in many patients without compromising outcomes.

Traditionally, women are offered chemotherapy and/or antihormonal therapy and/or targeted human epidermal growth factor receptor 2 (HER2) based on nodal status, tumor size, hormone receptor status, HER2 receptor status, and menopausal status. The type and duration are individualized by the patient's particular cancer.

Radiation

In patients undergoing breast conservation therapy, radiation is usually recommended to reduce local recurrence rates to 8–10 %. Studies have shown that in elderly patients with early stage node-negative cancers treated with lumpectomy and antihormone therapy, radiation may be omitted without compromising survival. Patients who undergo mastectomy are usually spared the need for radiation except in those with tumors >5 cm, T4 disease (invasion into skin or chest wall), or four or more positive lymph nodes. Recent studies suggest

that there may be a benefit in adding radiation in women with one to three lymph nodes positive.

Adjuvant Therapy

In general, for patients who are lymph node positive, there is a recommendation to receive chemotherapy, and this is usually an anthracycline- and taxane-based regimen. For patients who are hormone receptor and HER2 negative, there is a recommendation for chemotherapy regardless of their lymph node status. Patients who are HER2 positive, regardless of hormone and lymph node status, usually receive chemotherapy and HER2-targeted therapy (trastuzumab or lapatinib). Recently, the 21-gene RT-PCR assay has further impacted management decisions regarding additional chemotherapy. It is used to stratify patients who are hormone receptor positive, lymph node negative into different prognostic groups (low, intermediate, and high risk of recurrence) with the low-risk group deriving little benefit from chemotherapy and the high recurrence group being more likely to benefit. There are ongoing studies to determine the benefit of chemotherapy in the intermediate-risk group. Finally, in *premenopausal* patients who are hormone receptor positive, tamoxifen may be given for 5 years; in *postmenopausal* patients, an aromatase inhibitor will be given for 5 years. Current studies indicate there may a benefit to treating patients with antihormone therapy for longer than 5 years.

Stage IV and Inflammatory Breast Cancer

In patients with stage IV (metastatic) cancer, surgery has no role except for palliation.

Inflammatory breast cancer is a rare and aggressive form of breast cancer and constitutes 2–3 % of breast cancer cases in the United States each year. Clinical signs—including skin thickening and redness, peau d'orange, breast swelling, breast warmth, and nipple flattening or retraction—are usually seen in a rapid sequence of changes. A punch biopsy of the affected skin will often show dermal lymphatic invasion. Initial therapy is with induction chemotherapy, and if no evidence of metastatic disease, a modified radical mastectomy is performed and then radiation therapy. If the cancer is hormone positive, tamoxifen or an aromatase inhibitor is utilized.

64
Nipple Discharge

Faaiza T. Vaince and Andrea Madrigrano

Introduction

Nipple discharge can be multifactorial in origin. Most commonly, it is a benign finding secondary to metabolic imbalances, intraductal papillomas, duct ectasia, fibrocytic disease, or drug side effects. However, it may also be a sign of an underlying malignancy, and therefore it is imperative to have a systemic approach to its workup (Fig. 64.1).

History and Physical

The first step in working up a patient with nipple discharge is to obtain a thorough history and physical. Important questions should delineate the characteristics of the discharge (i.e., milky, purulent, serous, serosanguinous, or bloody). Serous, serosanguinous, or bloody nipple discharge is concerning for a malignant etiology. Purulent discharge suggests an infectious etiology. Milky discharge may be physiologic or related to hormonal imbalances.

Soliciting associated constitutional symptoms is necessary when obtaining the history. Visual changes may suggest a central nervous system process (pituitary lesion) that would affect prolactin levels. Anorexia, unintentional weight loss, or bone pain suggests a neoplastic process. Fevers and chills would suggest an infectious etiology. A careful review of the patient's medications may lead to a drug-related etiology.

On physical examination, it is important to correlate the patient's description of the discharge with your own findings. On exam, reproducible discharge may be unilateral or bilateral and may be from a single duct or multiple ducts. Attention should be given to the characteristics and location of any discharge that is expressed. A complete exam of the breast and axilla may reveal associated masses, skin changes, or lymphadenopathy that would raise concern for malignancy.

Spontaneous Nipple Discharge

Bilateral Spontaneous Discharge

Bilateral discharge, a multicentric problem, *is usually physiologic and benign*. Physiologic discharge can be related to hormonal imbalances, medications, or neurogenic stimulation. True galactorrhea is copious, spontaneous bilateral milky discharge in a non-lactating woman. Hormonal etiologies include hyperprolactinemia, pregnancy, and renal, adrenal, or thyroid disease. These can be diagnosed with the appropriate lab work, including a pregnancy test, renal panel, thyroid panel, and prolactin levels. Medications that cause increased prolactin levels with subsequent nipple discharge include antipsychotics, antidepressants, antiemetics, and antihypertensives. Neurogenic stimulation causing nipple discharge may be from excessive breast manipulation, poor-fitting undergarments, or a healing chest wound. If physiologic nipple discharge is identified, reassurance and treatment of the underlying etiology is necessary.

Unilateral Spontaneous Discharge

Unilateral spontaneous discharge warrants a more cautious evaluation. Spontaneous single-duct nipple discharge that is serous or sanguinous requires radiological and surgical evaluation. *Cytologic evaluation of the nipple discharge specimen has fallen out of favor as it rarely provides a definitive diagnosis that would change management*. Initial radiologic studies include an ultrasound and mammogram. If a dilated duct with an intraluminal mass is identified on imaging, the etiology is most likely an **intraductal papilloma**. Surgical excision is necessary to rule out malignancy. If a mass or cluster of microcalcifications is identified on imaging, an imaged-guided core biopsy can provide the diagnosis that would then guide further surgical and medical management.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_64, © Springer New York 2015

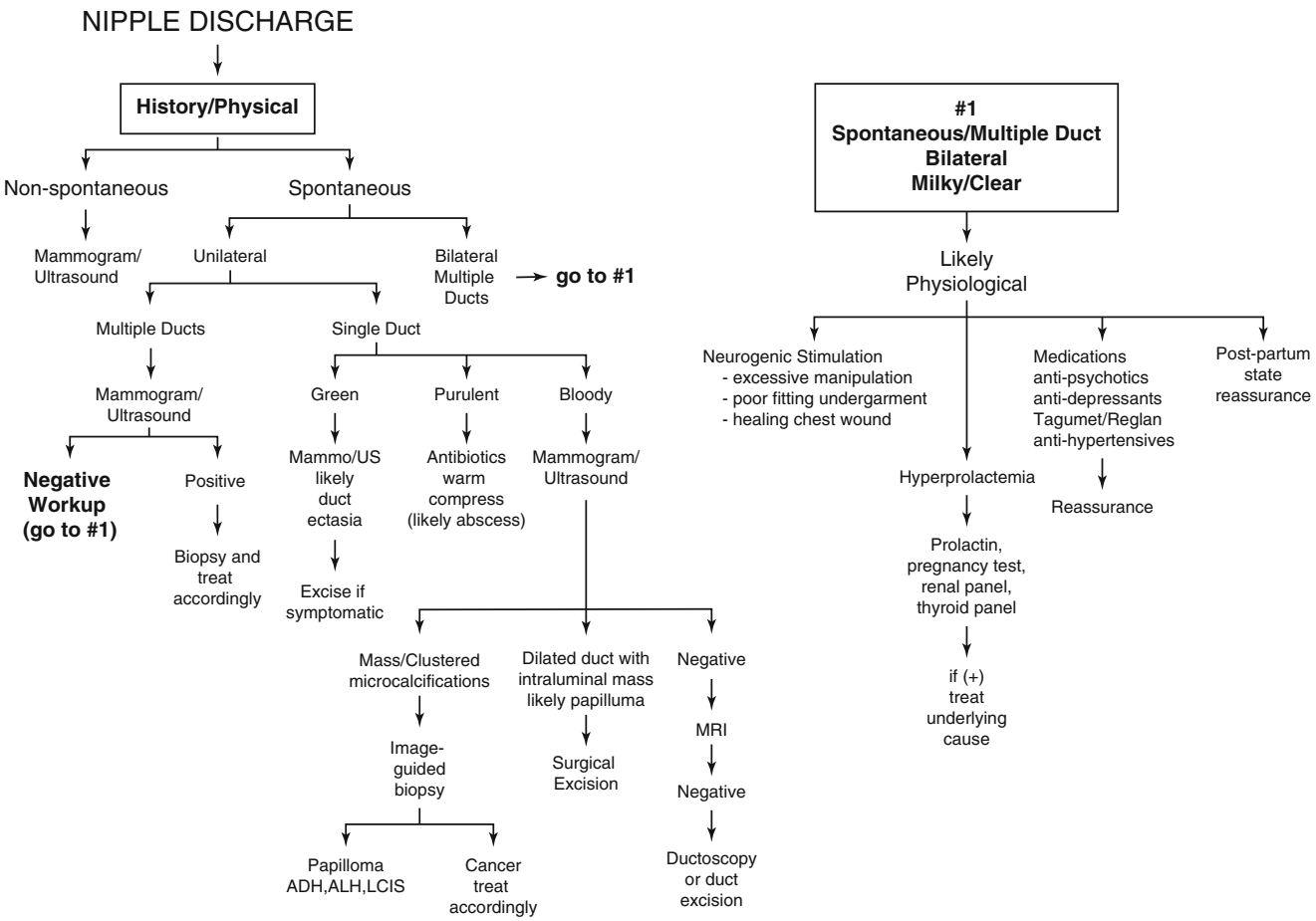

FIG. 64.1 Treatment algorithm for nipple discharge. *Mammo* mammogram, *US* ultrasound, *ADH* atypical ductal hyperplasia, *ALH* atypical lobular hyperplasia, *LCIS* lobular carcinoma in situ, *MRI* magnetic resonance imaging

If no finding is identified on initial radiological studies, an MRI may be indicated to help identify occult lesions. Ductoscopy and galactograms are sometimes utilized; however, they have recently fallen out of favor. If these too are negative, a terminal duct excision maybe necessary to definitively rule out neoplastic processes.

Single-duct, spontaneous nipple discharge that is purulent suggests an infectious etiology and may be related to an abscess. This should be treated with antibiotics, warm compresses, and aspiration as needed. Green-colored discharge is usually secondary to duct ectasia, which can be confirmed with a mammography or ultrasound. If such discharge is truly bothersome, excision of the area of pathology is indicated.

Nonspontaneous Nipple Discharge

Nonspontaneous discharge is usually related to an inciting or temporal factor that can be elicited when taking a history. Additional workup should include imaging studies, which would again be a mammogram or ultrasound. If these studies are negative, then reassurance and clinical follow-up is appropriate. If pathology is identified, then a diagnostic biopsy is indicated, which then guides further treatment.

65
Ductal Carcinoma In Situ

Anna B. Katz

Epidemiology and Risk Factors

Ductal carcinoma in situ (DCIS) affects 32.5 per 100,000 women in the US, most commonly affecting those in their sixth decade of life. The risk of developing DCIS increases with increasing age. DCIS rarely occurs in women younger than age 30. DCIS, a precursor to invasive carcinoma, is the proliferation of malignant ductal epithelial cells without microscopic invasion through the basement membrane. *DCIS usually presents as clusters of microcalcifications on mammogram* (90 % of DCIS cases) but less commonly may present as a palpable mass (Fig. 65.1). Since the 1970s, the incidence of DCIS has increased to 20–25 % of newly diagnosed breast cancers in the USA, likely because of screening mammography. Up to 90 % of DCIS is diagnosed because of mammographic abnormalities. While DCIS is a significant disease, the 10-year survival rate approaches 100 %.

There are five subtypes of DCIS, which include: comedo, cribriform, solid, papillary, and micropapillary. Of these subtypes, *comedo is the most aggressive, acts most similarly to invasive cancers, and has the worst prognosis*. Comedo lesions are poorly differentiated, high-grade lesions that are characterized by central ductal necrosis, often with microcalcifications within the area of necrosis. They have marked atypia, higher rates of mitosis and are associated with larger tumors with increased incidence of multicentricity and microinvasion. The other four subtypes are less aggressive with lower cytologic grade and lower rates of mitosis. Along with grade, the most important factors to consider in the treatment of DCIS are extent of disease and margin status. While the natural history of DCIS is not clearly known, it is believed that most will progress to invasive disease over time.

Risk factors for developing DCIS and invasive breast cancers include: age (increases with increasing age), exposure to unopposed estrogen (younger age at menarche and later age of menopause), increased age at first live birth (after age 30), parity, exogenous hormone use, family history of breast cancer, female gender, personal history of breast cancer, obesity, and moderate to severe levels of alcohol use. There are certain breast lesions that also carry an increased risk of developing breast cancer, which include proliferative breast disease (raises risk 1.5–2 times that of normal), atypical ductal hyperplasia and atypical lobular hyperplasia (raises the risk 3.5–5 times), and lobular carcinoma in situ (raises the risk 7–11 times).

Diagnosis

A diagnosis of DCIS requires tissue. The standard of care is to perform a **core** needle biopsy for any concerning lesion seen on mammogram and ultrasound or palpated on clinical exam. If this is not diagnostic, not concordant, or reveals atypia or another high-risk lesion, the next step is excisional biopsy. Incisional biopsies are not commonly performed. Fine needle aspiration (FNA) is often used to biopsy lesions that are palpable or seen on ultrasound, but *FNA cannot reliably distinguish between DCIS and invasive cancer*. Lesions found on physical exam are often more advanced tumors and are more likely to have occult invasion and multicentricity. The typical mammographic finding of DCIS are clusters of microcalcifications, which represent dystrophic calcium deposits from necrotic cells. A less common mammographic finding for DCIS is a soft-tissue density. A negative core needle biopsy in the face of a suspicious mammogram does not rule out DCIS or invasive breast cancer; this phenomenon is known as **discordance**. If pathology is discordant with imaging and/or clinical findings, it is imperative to proceed with excisional biopsy. This may necessitate needle localization if the lesion is nonpalpable and seen only on mammogram.

Treatment

The first step in treating DCIS is surgical, consisting of either breast conservation (lumpectomy) or total mastectomy, depending on extent of disease and multicentricity. All patients having a total mastectomy should undergo

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_65, © Springer New York 2015

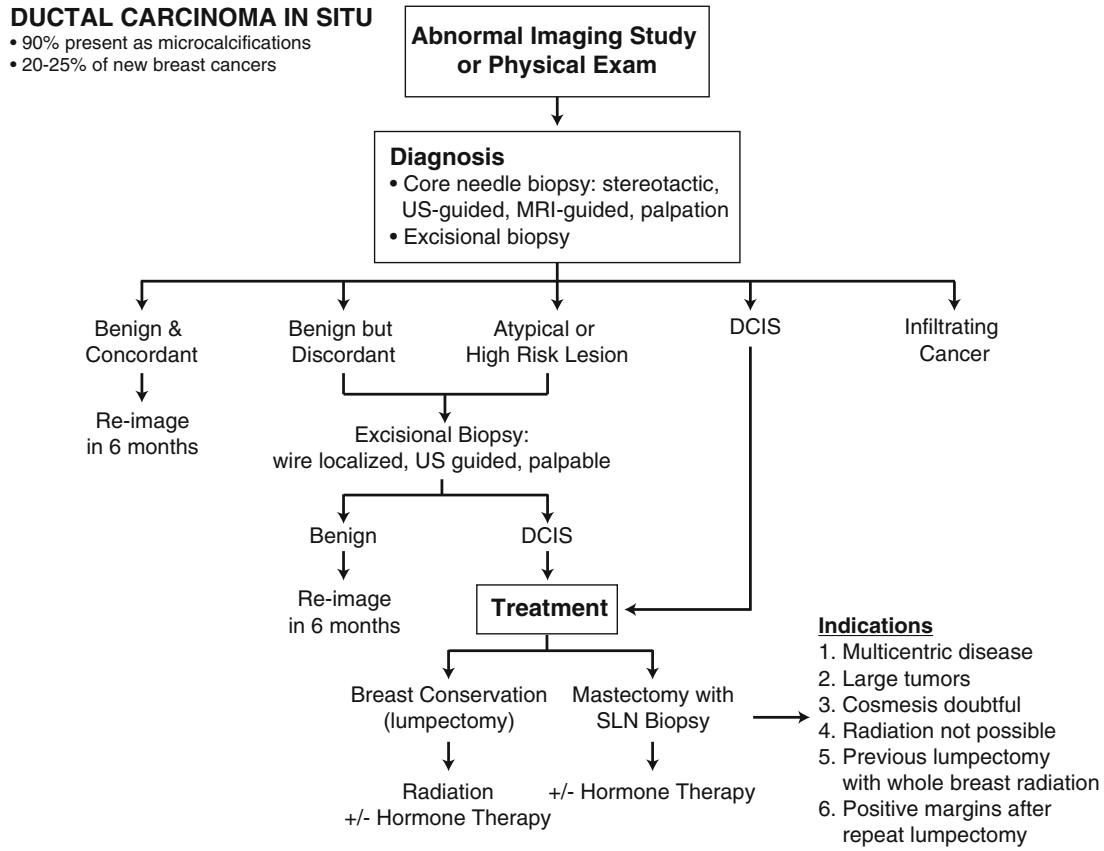

FIG. 65.1 Treatment algorithm for ductal carcinoma in situ (DCIS). *US* ultrasound, *MRI* magnetic resonance imaging

sentinel lymph node (SLN) biopsy as some patients will have invasive disease found on final pathology. Indications for mastectomy include multicentric disease, large tumors or smaller tumors in smaller breasts that would prevent an acceptable cosmetic result with breast conserving therapy (BCT), contraindications to radiation, previous BCT followed by whole breast irradiation, diffuse malignant-appearing calcifications throughout the breast, or failed attempts at BCT (continued positive margins despite re-excision). The definition of a clear margin has historically been controversial; some centers accepting a 1–2 mm margin versus other centers requiring up to 10 mm margin for DCIS. However recently, the society of surgical oncology (SSO) and americal society for radiation oncology (ASTRO) released a consensus statement saying that no tumor at inked margin is an adequate margin. Women treated with lumpectomy have equivalent survival as those treated with mastectomy.

The second component to treatment is the addition of postoperative radiation, either in the form of whole breast irradiation or partial breast irradiation. Radiation is generally recommended to all patients who undergo BCT. Some patients may not have radiation depending on their age and comorbidities, in which case the risks and benefits of adjuvant radiation are weighed. Radiation may lower the risk of local recurrence in patients undergoing BCT by up to 50 %. Finally, while patients with DCIS do not require chemotherapy, most will get endocrine therapy in the form of Tamoxifen for 5 years depending on the estrogen receptor status of their tumor.

Sentinel lymph node (SLN) biopsy and axillary lymph node dissection (ALND) are generally not recommended for patients with pure DCIS. ALND is a potentially morbid procedure and the risk of lymphedema is as high as 30 %. As previously mentioned, SLN biopsy is recommended for women undergoing mastectomy and may be recommended for women undergoing lumpectomy with more aggressive pathology on core needle biopsy, such as comedo necrosis.

Follow-up mammography is recommended at 6-month intervals for the first 2 years after surgery and then at yearly intervals. Clinical breast exams are performed biannually for the first 5 years and then annually.

66
Genetic Predisposition to Breast Cancer

Mehra Golshan

Introduction

Breast cancer is the most common cancer in women and the second leading cause of cancer-related deaths. In 2011, approximately 232,000 American women were diagnosed with invasive breast cancer and 40,000 died from the disease; one in 210 breast cancer cases occur in women under 40.

BRCA1 and BRCA2

Inherited predisposition to breast and ovarian cancer accounts for 5–10 % of all breast cancer cases diagnosed each year in the United States. Mutations in the BRCA1 and BRCA2 genes are thought to account for the largest proportion of hereditary breast cancers and explain about 10 % of breast cancer cases in women diagnosed under the age of 35 (Fig. 66.1). The lifetime risk of breast cancer for BRCA1 or 2 mutation carriers is estimated by several studies to be between 45 % and 80 %.

In 1990, genetic studies provided initial evidence that connected the risk of early-onset breast cancer in some families to chromosome 17q21. Soon after, linkage to this same locus was established in families with hereditary breast and ovarian cancer syndrome. Miki et al. subsequently cloned the BRCA1 gene on chromosome 17q21 in 1994. Since its discovery, BRCA1 has been intensely researched. Analyses of thousands of samples in research and clinical contexts have identified many different germline mutations.

BRCA2 was cloned in 1995 after chromosome 13q12-13 was linked to families with both early-onset breast cancer and male breast cancer. BRCA2 contributes to fewer cases of early-onset breast cancer in the United States when compared to BRCA1 but confers an estimated 6–10 % lifetime risk of breast cancer in male mutation carriers. Germline mutations in BRCA2 are associated with increased risk of several other cancers in addition to breast and ovarian cancer, including cancers of the prostate, pancreas, stomach, gallbladder, and bile duct, as well as malignant melanoma.

BRCA3 has yet to be identified; however, a growing list of genes have been shown to confer either substantial increases in breast cancer risk in small numbers of patients or small increases in risk in larger population groups. These genes are currently being studied such as ATM, p53, CHEK2, PTEN and PALB2.

BRCA1 and BRCA2 are **tumor suppressor genes** that are involved in multiple cellular processes including DNA repair and transcriptional regulation in response to DNA damage, chromosomal stability, and cell-cycle regulation. Unlike classical tumor suppressor genes, however, *mutations in BRCA1 and BRCA2 are almost never seen in sporadic breast cancers*.

BRCA-Related Breast Cancer

BRCA1- and BRCA2-associated breast cancers differ from sporadic breast cancers in both clinical and pathologic characteristics. Mutation carriers have a younger age of cancer onset, with tumors occurring on average one decade earlier than those in non-mutation carriers. Furthermore, BRCA-related lesions possess worse histologic factors than sporadic tumors. Studies have demonstrated a greater degree of aneuploidy, higher nuclear and histologic grade, and higher proliferation indices in BRCA1-related lesions. BRCA1 tumors are also more frequently estrogen receptor (ER) and progesterone receptor (PR) negative than BRCA2-associated and nonhereditary breast cancers. BRCA2 tumors are predominantly high-grade invasive ductal carcinomas. Despite their high histologic grade, BRCA2 tumors demonstrate a luminal phenotype, and nearly 90 % are ER and/or PR positive. Compared to sporadic cancers, both BRCA1 and BRCA2 tumors are less often human epidermal growth factor receptor-2 (HER2) positive.

Although BRCA-linked breast cancers are associated with worse histological features and reduced levels of estrogen and progesterone receptor expression, survival analyses have been controversial. Most studies have found survival

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_66, © Springer New York 2015

GENETIC PREDISPOSITION TO BREAST CANCER

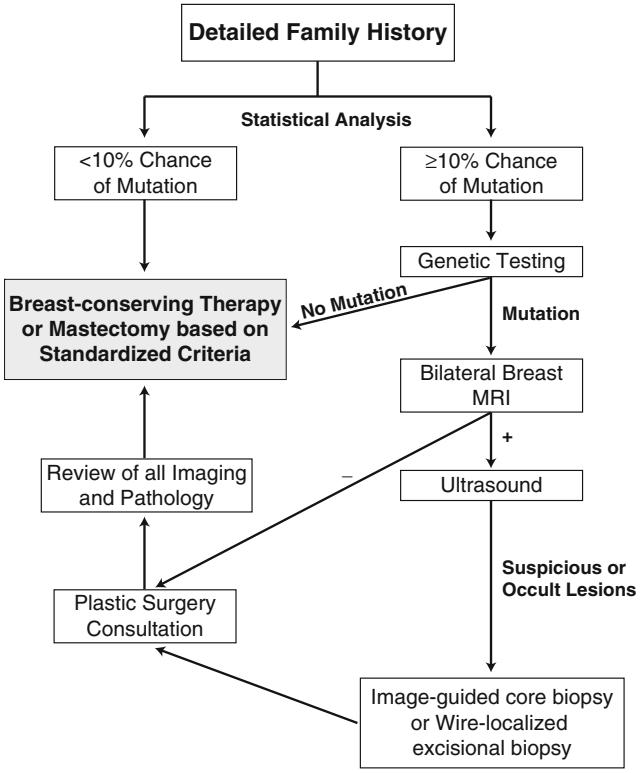

FIG. 66.1 Algorithm for surgical decision making for diagnosed breast cancer. *MRI* magnetic resonance imaging

rates in BRCA patients to be equivalent to those in patients with sporadic breast cancer. However, a few studies have demonstrated reduced survival in BRCA carriers, and two analyses showed BRCA1 mutation to be an independent negative prognostic factor in breast cancer. Unfortunately, many of these studies did not control for adjuvant therapeutic intervention or eliminate age as a confounding factor. In addition, most studies addressed BRCA1- and BRCA2-related cancers together rather than as two separate entities.

Studies addressing local recurrence in BRCA patients have also been inconsistent. Though the majority of reviews demonstrate no significant difference between genetic and sporadic cases, a few have shown increased ipsilateral breast tumor recurrence in BRCA patients. Most studies reporting an increased recurrence rate were small in number and none addressed the issue of adjuvant therapy. Furthermore, the increase in local recurrence did not adversely affect overall survival. In contrast, BRCA carriers do appear to have a higher probability of developing ipsilateral new primaries.

Studies looking at contralateral breast cancer risk have consistently shown a higher incidence of tumors in BRCA patients than in the general population. The average risk of developing a contralateral breast cancer ranges from 0.5 % to 0.8 % per year in the sporadic patient. This number increases to 2–3 % per year in a woman with an inherited predisposition to carcinoma based on BRCA1 or 2 mutation.

Women with an inherited predisposition to breast and ovarian cancer without a cancer diagnosis will often choose prophylactic mastectomy in the skin or nipple-sparing fashion with immediate reconstruction. Once a cancer diagnosis is made, most patients with a BRCA1 or 2 mutation will choose bilateral mastectomy with immediate reconstruction and appropriate lymph node evaluation. Consideration for breast conserving therapy especially in older BRCA 2 mutation carriers may be made with the caveat of higher new primary in the ipsilateral breast and contralateral breast, although local recurrence for the known breast cancer remains similar to those with sporadic disease.

67
Melanoma

Keith C. Hood and Steven Bines

Epidemiology

The incidence of melanoma is rising faster than any other malignancy and is the sixth most common cancer in North America. Melanoma accounts for only 4–5 % of all skin cancers but causes a majority of the deaths from skin malignancies. It is estimated that melanoma develops 20 times more frequently in whites than in blacks, and it occurs more commonly in the lower extremities in women and more often on the trunk and head and neck in men. The median age at diagnosis is 45–55 years old. The lifetime probability of melanoma is 1 in 57 for males and 1 in 81 for females. It is well established that exposure to sunlight, specifically solar ultraviolet (UV) radiation, increases the risk of developing melanoma. People incurring severe sunburns in childhood appear to be at a higher risk for melanoma years later. Other risk factors include a positive family history, Type 1 and Type 2 skin, dysplastic nevus syndrome, xeroderma pigmentosum, an increased number of common nevi, and one or more atypical nevi. People with more than 50 moles have a two- to threefold increase risk of melanoma.

Presentation

Cutaneous melanoma typically presents as a flat lesion that spreads over the surface of the skin and later becomes elevated (Fig. 67.1). If the lesion is allowed to progress, itching, bleeding, and ulceration will occur. The clinical features of melanoma are summarized in the mnemonic "**ABCDE**." Suspicious pigmented lesions are typically ***a***symmetrical, have changing irregular ***b***orders, have ***c***olor variation within the lesion, show an increase in ***d***iameter (>5 mm), and have an ***e***levated or ***e***volving surface. *All suspicious pigmented lesions require a biopsy and a formal clinical skin evaluation including an examination of the regional lymph nodes*.

Diagnosis

The most important prognostic indicator for melanoma is tumor thickness. Whenever possible, complete excisional biopsy should be performed to allow for pathologic examination of the entire lesion. The specific method of biopsy depends on the size of the lesion and its anatomic location. Regardless of method, biopsy specimens are full-thickness into the subcutaneous tissue. For small lesions, an excisional biopsy including a margin of 1–3 mm of normal skin is performed. When an incisional biopsy of a large lesion is done, at least one punch is placed through the most elevated portion to accurately classify its thickness. This method is also acceptable in lesions where an excisional biopsy would be impractical (the face, palm, sole, etc.)

The pathology report should contain information regarding tumor thickness, the presence or absence of ulceration, and mitotic index. In 1969, **Clark** and associates described a classification of melanoma based on the extent of tumor invasion relative to the anatomic layers of the skin. Clark's level of invasion was shown to correlate with survival. In 1970, **Breslow** described a more straightforward system based on measuring the vertical thickness of the tumor in millimeters. This method was found to be accurately reproducible between pathologists and had a strong correlation with 5-year survival. The prognosis worsens with increasing thickness as a continuous logarithmic function. **For this reason, the Breslow thickness is considered more prognostic**. For each tumor thickness, the presence of ulceration is associated with a worsened prognosis. The mitotic rate was incorporated into the 2010 tumor, node, metastasis (TNM) staging system based upon the observation that it was the second most important prognostic factor for localized melanoma. The area of dermis containing the most mitoses (the "hot spot") is identified. Once this is identified, mitoses in adjacent fields within a total area of 1 mm^2 are counted to determine the mitotic rate. A higher mitotic index significantly correlates with declining survival rates.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_67, © Springer New York 2015

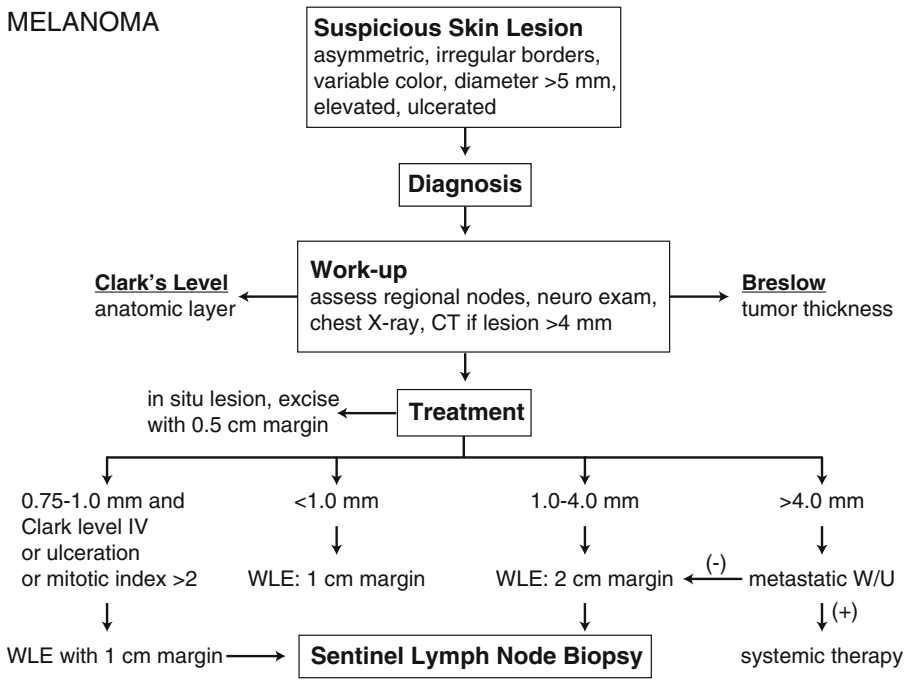

FIG. 67.1 Melanoma treatment algorithm. *CT* computed tomography, *WLE* wide local excision

There are four major subtypes of invasive cutaneous melanoma: superficial spreading, nodular, lentigo maligna, and acral lentiginous. Superficial spreading melanoma is the most common subtype, accounting for approximately 70 % of all melanomas. More than 60 % of superficial spreading melanomas are diagnosed as thin, highly curable tumors of less than 1-mm thickness. Nodular melanomas are the second most common type, accounting for 15–30 % of all melanomas, and are characterized by an early vertical growth phase. As a result, more than 50 % of melanoma lesions greater than 2 mm in thickness are nodular. Lentigo maligna accounts for 10–15 % of all melanomas. The acral lentiginous subtype accounts for less than 5 % of all melanomas. However, it is the most common type of malignant melanoma among dark-skinned individuals. Acral lentiginous melanomas arise most commonly on palmar, plantar, and subungual surfaces.

Workup

Patients with cutaneous melanoma should undergo a complete physical examination, with emphasis on the skin and regional lymph nodes. This should also include a screening neurologic examination. All patients should undergo a baseline chest X-ray and serum lactate dehydrogenase (LDH). Additional metastatic workup is not indicated unless the patient has specific symptoms or findings suggestive of metastatic disease. For patients with melanomas greater than 4-mm thick or findings of metastatic disease, the workup should include MRI of the head, CT of the chest, abdomen and pelvis, and a positron emission tomography (PET) scan.

Invasive melanomas are capable of recurring locally, metastasizing to the regional lymph nodes or developing distant metastases. Both satellite (<2 cm from the primary tumor) and in-transit (>2 cm from the primary tumor) metastases may develop along the route of spread from the primary tumor to the regional lymph nodes. These lesions are incorporated in the N (node) parameter of the TNM staging system.

Treatment

All melanomas are treated locally with wide local excision (WLE). This excision is full-thickness and carried down to, but not through, the underlying fascia. In most cases, closure with elevation of local advancement flaps is possible. Occasionally, skin grafts or more complex flaps may be required. Several large clinical trials have studied the optimal surgical resection margin for primary melanomas. For in situ lesions, the margin of excision should be 0.5 cm. For invasive melanomas up to 1 mm in thickness, a wide local excision with a 1 cm margin is indicated. For lesions between 1 and 2 mm thickness, a margin of 2 cm is appropriate. For melanomas between 2 and 4 mm thick as well as those greater than 4 mm, clinical trials have not demonstrated a benefit of excision margins greater than 2 cm; therefore a 2 cm margin is recommended. *On the extremities, these excisions should be performed along the longitudinal axis to capture the draining lymphatics to the regional lymph nodes.*

It has been shown in several prospective, randomized controlled trials that prophylactic lymph node dissection does not confer a survival advantage. Patients with clinically positive biopsy-proven lymph node metastases should undergo the appropriate radical lymphadenectomy, an anatomically defined complete lymph node dissection. The number of metastatic lymph nodes is a significant prognostic factor; patients with only one positive node have a better prognosis than those with more than one positive node.

Sentinel Lymph Node Biopsy (SLN)

For patients with invasive melanomas 1 mm or thicker, or patients with melanomas 0.75- to 1-mm thick associated with Clark's level 4 depth of invasion, ulceration, and/or a mitotic index greater than 2, a sentinel lymph node biopsy (SNLB) is indicated. This is based upon the concept that cutaneous melanoma has specific patterns of lymphatic spread and that one or more nodes are the first to be involved with metastatic disease within a given lymph node basin. If the sentinel lymph nodes are not involved, the entire basin should be free of tumor. Therefore, lymphatic mapping with SNLB permits the identification of patients with positive nodes and avoids lymph node dissection in those without nodal involvement.

Technetium-labeled dye (Tc-99) can be used for both preoperative mapping and for sentinel node identification in the operating room using a handheld gamma probe. Further confirmation can be obtained by injecting vital blue dye (isocyanine blue) in the dermis at the primary tumor site approximately 15 min before SLNB. In addition to the sentinel node(s) identified via radioactive and/or blue dye, enlarged or acanthotic nodes are also removed.

Immunohistochemical markers (HMB45, S100, MART-1, and Melan-A) and hematoxylin and eosin (H&E) staining are performed on the thin sections of the sentinel node for diagnostic purposes. Completion radical lymphadenectomy is indicated for patients with tumor involvement of the sentinel lymph node. However, a small number of sentinel nodes will be falsely reported as negative even with optimal technique. Approximately 3 % of patients with a negative SLNB subsequently relapse in the regional lymph nodes and require a radical lymphadenectomy.

The Multicenter Selective Lymphadenectomy Trial (MSLT-1) is the largest trial to address the role of lymphatic mapping with SNLB in determining prognosis and its impact on survival. Patients with intermediate thickness (1.2–3.5 mm) primary melanomas randomly assigned to wide local excision plus SLNB with immediate completion lymphadenectomy for positive nodal status had longer disease-free survival, lower relapse rates, and improved overall survival compared with patients assigned to wide local excision plus observation with completion lymphadenectomy at the time of development of clinically evident lymph node disease.

Patients with a positive sentinel node should undergo a metastatic workup prior to regional lymphadenectomy if done in a staged manner. Those with positive sentinel nodes as well as patients with tumors greater than 4-mm thick without ulceration or tumors 2–4-mm thick with ulceration have been the primary focus of studies evaluating the efficacy of adjuvant therapy. The most promising results have been reported with high-dose interferon alpha. Initial studies with interferon alpha showed a statistically significant improvement in overall and disease-free survival. Subsequent trials have confirmed the improvement in disease-free survival but failed to demonstrate an improvement in overall survival.

There is no standard treatment for metastatic disease. Solitary metastases should be evaluated for resection when anatomically feasible, the patient is otherwise healthy, and there is no evidence of additional metastatic disease over a short period of observation.

For patients rendered disease-free after surgery, follow-up should include careful physical examination every 3 months for the first 2 years, every 6 months for the next 3 years, and annually thereafter. The patient should undergo an annual chest X-ray and a serum LDH every visit. Serum LDH is sometimes elevated in patients with recurrent or metastatic disease and is considered helpful but is not highly sensitive or specific.

68
Solitary Pulmonary Nodules

David D. Shersher and Anthony W. Kim

Introduction

There is no universally accepted definition for pulmonary nodules, but they typically are defined as solitary, well-circumscribed lesions that are surrounded by normal lung parenchyma, not adjacent to hilum or mediastinum, and not associated with atelectasis or pleural effusion (Fig. 68.1). These lesions usually are less than or equal to 3 cm in size; larger lesions are termed solitary masses and are highly suspicious for malignancy. Lesions that have significant ground glass opacity are not considered solitary pulmonary nodules (SPNs).

Patients with SPNs are often asymptomatic, and the nodules are usually found incidentally on chest imaging. It is estimated that greater than 150,000 SPNs are diagnosed by chest roentography or computed tomography (CT) annually. With recent evidence demonstrating a benefit to screening CT scans, the incidence may even be greater.

The differential diagnosis of SPNs includes both benign and malignant processes. The majority of these lesions are caused by nonspecific or infectious granulomas associated with aspergillosis, coccidiomycosis, cryptococcus, histoplasmosis, or tuberculosis. The second most common cause of benign SPNs is hamartomas. SPNs that harbor malignancy often are associated with adenocarcinoma, followed by squamous cell carcinoma and, less frequently, metastasis. Carcinoids and small cell cancers causing SPNs are much less common but should be included in the differential.

Evaluation

Any pulmonary nodule that is greater than 3 cm is considered malignant until proven otherwise. For lesions smaller than 3 cm, the Veterans Affair and Mayo Clinic models can be used to "assign" risk of malignancy. *Low-risk characteristics* include age less than 40 years, no smoking history, no history of prior malignancy, a doubling time less than 1 month or greater than 1 year, and a benign morphological pattern (smooth, dense, and solid nodules with associated homogenous calcifications). *High-risk characteristics* include age greater than 40 years, a smoking history, a history of prior malignancy, a doubling time between 1 month and 1 year, and suspicious morphological pattern (irregularly contoured, non-solid nodules with heterogeneous or absent calcifications).

In general, the Fleischner criteria should be applied to SPNs. For *lesions less than 8 mm in size*, *low-risk patients* should undergo repeat imaging based on size; SPNs up to 4 mm warrant no follow-up, between 4 and 6 mm require reimaging in 1 year, and 6–8 mm necessitate reimaging in 6 months. For *high-risk patients*, SPNs up to 4 mm warrant reimaging in 1 year, between 4 and 6 mm require reimaging in 6 months, and 6–8 mm necessitate reimaging in 3 months. *Lesions larger than 8 mm* should be evaluated more closely based on cancer risk. Low-risk patients require radiographic follow-up every 3 months, while intermediate- and high-risk patients require tissue biopsy and/or positron emission tomography (PET) – although the guidelines for action with this modality are less defined.

With increased use of high-resolution, low-dose computed tomography, all patients with serially followed SPNs should undergo this type of imaging rather than chest roentography for comparative follow-up.

Biopsy and Surgery

If a critical review of the SPN on imaging warrants a tissue diagnosis, a variety of methods can be used. Classically, percutaneous biopsies have been used for noncentral nodules and transbronchial biopsies for central nodules. More sophisticated transbronchial biopsy techniques using radial probe endobronchial ultrasound technology with or without electromagnetic navigational bronchoscopy can be used to access more peripheral nodules.

The popularization of video-assisted thoracoscopic surgery (VATS) has also facilitated the sampling of suspicious nodules through better lesion visualization, sampling of the entire nodule, and management of post-procedure pneumo- or

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_68, © Springer New York 2015

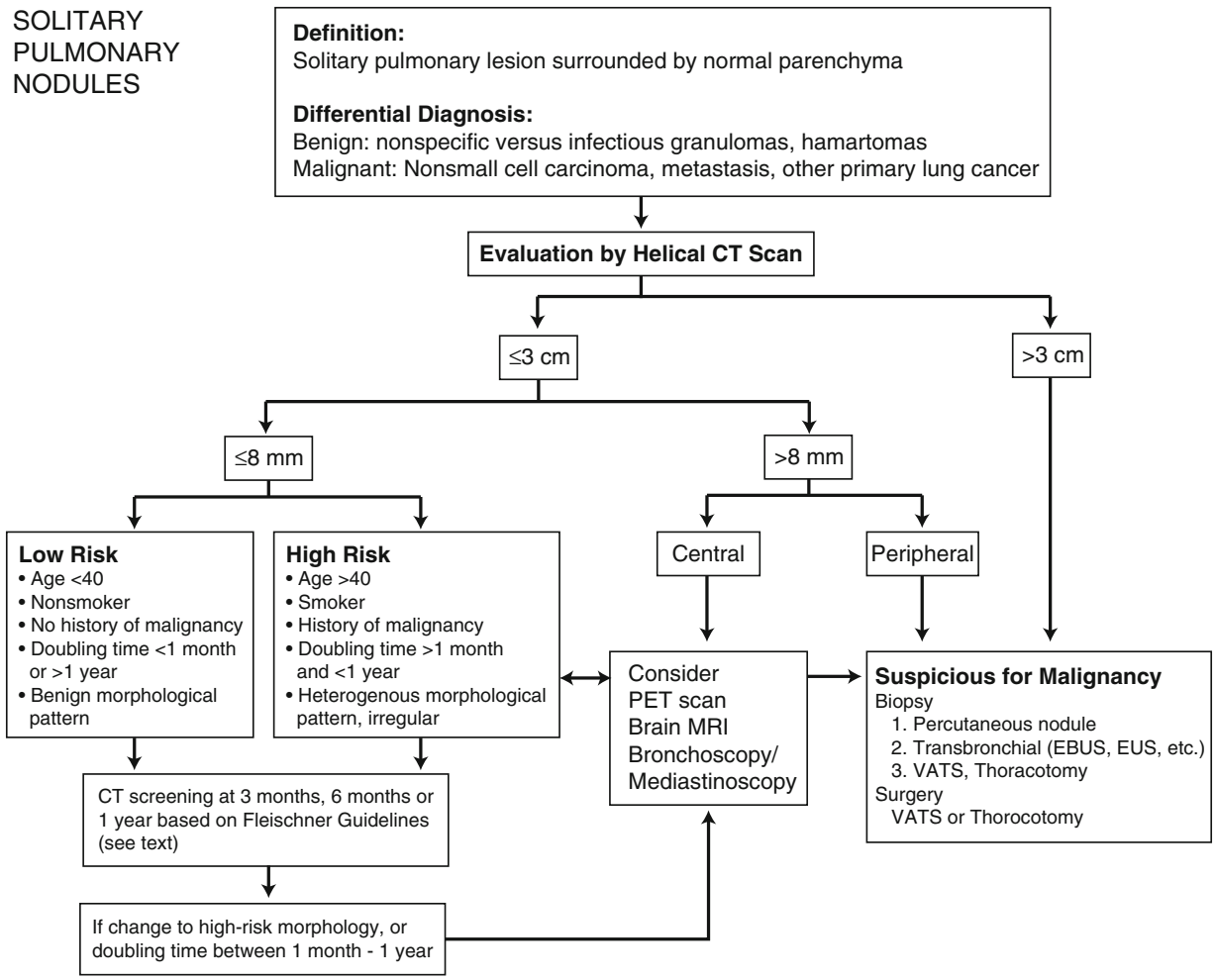

FIG. 68.1 Treatment algorithm for solitary pulmonary nodules. *CT* computed tomography, *PET* positron emission tomography, *MRI* magnetic resonance imaging, *VATS* video-assisted thoracoscopic surgery, *EBUS* endobronchial ultrasound, *EUS* endoscopic ultrasound

hemothorax. An added benefit of a VATS wedge resection is that the diagnostic procedure can immediately precede a therapeutic formal anatomic resection in the same general anesthetic setting. Deeper lesions may be accessible by the VATS approach through adjuncts such as needle localization.

Patients scheduled to undergo VATS for biopsy or for definitive resection require preoperative surgical workup, which includes pulmonary function tests. Patients at high risk for complications after thoracotomy usually have a preoperative FVC <30 % of predicted or a FEV1 of <1 L or a predicted postoperative FEV1 of less than 800 mL. Perfusion imaging can be used as an adjunct to better determine preoperative pulmonary risk.

Conclusion

Most SPNs are found incidentally during imaging for other reasons and lead to often unnecessary, expensive, and potentially morbid workup unless established guidelines are followed. Although the majority of these SPNs will harbor no malignancy, the potential of cancer is real; and with a rising incidence of non-small cell lung cancer predicted to surpass 250,000 per year, appropriate stepwise diagnosis and surveillance is critical to reduce overall morbidity and mortality from this disease process.

69
Evaluation of the Suspicious Neck Mass

Marissa Le

Introduction

Neck masses can be classified as congenital, inflammatory, or neoplastic. The history and physical exam provide key information regarding the most likely etiology and guide further workup of the mass (see Table 69.1 and Fig. 69.1). History should include tobacco and alcohol use, infectious exposure, and travel. Inflammatory masses are the most common cause among children and young adults. Congenital lesions become clinically apparent before the age of 30 but cannot be excluded in older patients. For patients over 40 years of age, malignancy should be at the top of the differential as 85 % of neck masses are neoplastic, and most of those are malignant.

Congenital Neck Masses

Branchial cleft cysts account for 17 % of pediatric congenital neck masses. Branchial cleft cysts arise from any of the branchial arches and are numbered accordingly. Remnants of the branchial cleft become apparent when they fill with fluid and become secondarily infected and may drain through a cutaneous sinus or fistula tract. The typical patient is a child with a lateral neck mass that fluctuates in size with viral illnesses. First branchial cleft cysts are rare and are intimately associated with the external auditory canal, the parotid gland, and the facial nerve, as such are difficult to resect completely. Second branchial cleft cysts are the most common and form a cystic mass in the mid or lower neck anterior to the sternocleidomastoid muscle. A fistula tract may run from the skin between the internal and external carotid arteries and open internally into the ipsilateral tonsillar fossa. Third branchial cleft cysts are lower on the neck, anterior to the sternocleidomastoid. The internal site of the fistulous tract drains into the pyriform sinus. Treatment is elective surgical excision. Preferred treatment of an actively infected branchial cleft cyst is antibiotics, with complete surgical excision 3–5 weeks later.

Thyroglossal duct cysts are the most common congenital lesion, comprising one-third of all pediatric congenital lesions. They are remnants of the thyroglossal duct formed by the descent of the thyroid diverticulum from the foramen cecum at the base of the tongue into the neck. These cysts are found in the midline anywhere along the course of the duct from the foramen cecum to the suprasternal notch. Similar to branchial cleft cysts, they fluctuate in size and can become secondarily infected. On exam, these *midline lesions elevate with swallowing and protrusion of the tongue*. Surgical excision with the Sistrunk procedure entails the removal of the cyst, the tract, the central portion of the hyoid bone, and a cuff of tissue at the tongue base. The Sistrunk procedure has a 10 % recurrence rate compared to 50 % recurrence with simple excision. Prior to excision, ultrasound or thyroid scan should be performed to ensure the presence of a functioning thyroid tissue as the thyroglossal duct cyst may contain all the thyroid tissue and complete excision will render the patient permanently hypothyroid.

Lymphatic malformation is a congenital mass of dilated lymphatic channels that lacks communication to normal drainage pathways. These masses may be microcystic or macrocystic (cystic hygroma), multilobulated, and multiloculated. On exam, they are soft, smooth, nontender, poorly circumscribed masses that are compressible and can be transilluminated. These malformations may fluctuate in size with infection and intralesional hemorrhage and may compromise the child's airway. Most cause only a cosmetic defect. Diagnosis is confirmed with computed tomography (CT) or magnetic resonance imaging (MRI). Treatment may be observation, surgical excision (which is technically difficult and often incomplete), or intralesional injection of sclerosing agents, which has met with mixed success.

Hemangiomas have a very characteristic appearance: bluish coloration, warmth, easy compressibility with rapid refilling, and occasionally a bruit. Typically hemangiomas undergo a growth phase early on, followed involution at age 18–24 months with watchful waiting. Most resolve with watchful waiting.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_69, © Springer New York 2015

TABLE 69.1. Main classifications of neck masses.

	Inflammatory	Congenital	Neoplastic
Age	Any age, more commonly <40 years old	Less than 30 years old	>40 years old
History	Acute onset, immunocompromised	Present at birth, fluctuates over time	Slow growth Tobacco and alcohol use Otalgia, oral pain, voice changes
Physical exam characteristics	Rapid growth, tender, warm, erythema, fluctuant or small, rubbery, mobile	Soft, ballotable, mobile, cystic. Draining cutaneous tract	Firm, fixed, nontender, cranial nerve palsy
Systemic symptoms	Rash or skin lesion, high fevers, night sweats, leukocytosis, splenomegaly	Only when acutely infected	Low-grade fever, weight loss, night sweats

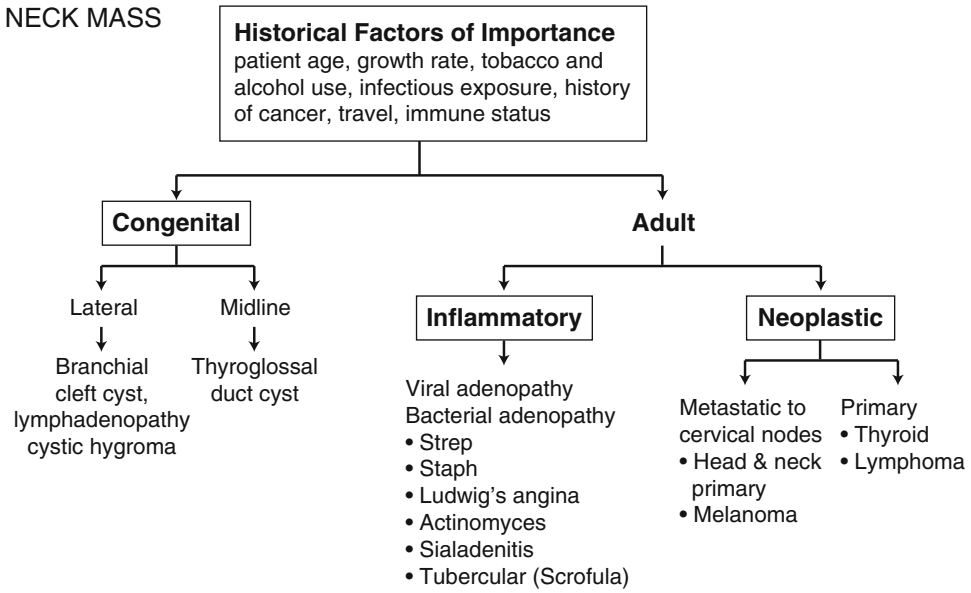

FIG. 69.1 Algorithm for evaluation of suspicious neck mass

Inflammatory Neck Masses

Viral reactive lymphadenopathy is a common cause of infectious neck masses. Examination will reveal multiple soft, mobile, small (<2 cm) lymph nodes. Epstein–Barr virus causes mononucleosis, characterized by nonspecific symptoms including fever, malaise, exudative pharyngitis, splenomegaly, and increased atypical white blood cell count. Patients typically present with diffuse posterior cervical lymphadenopathy. Diagnosis is confirmed with a monospot test, which may be falsely negative early in the illness. Treatment is supportive care. Adenopathy typically resolves in 4–6 weeks.

Acute bacterial lymphadenitis is the most frequent cause of a infectious mass in the neck. It is inflammatory enlargement of one or more lymph nodes usually secondary to a staphylococcal or group A beta-streptococcus infection of the upper respiratory tract, oral cavity, or skin. On exam, the nodes are tender, mobile, and occasionally with overlying erythema. Necrotic lymph nodes will be fluctuant on exam. Ultrasound can differentiate soft tissue induration from a fluid-filled mass, but CT is diagnostic. Treatment is antibiotics and incision and drainage if the node is fluctuant. Initial antibiotics selection should cover methicillin-resistant *Staphylococcus aureus* (MRSA) until initial culture results reveal the susceptibility of the causative organism.

Ludwig's angina is a life-threatening dental infection originating in one of the posterior mandibular molars. Infection spreads into the submandibular space. Induration and edema in this limited space displace the tongue posteriorly into the oropharynx and can quickly threaten the airway. The patient appears toxic and cannot swallow oral secretions, and examination reveals firm edema of the floor of the mouth and the submandibular space. *The patient's airway should be secured immediately*. Treatment is surgical drainage of the submandibular space, intravenous (IV) antibiotics, and maintenance of airway.

Actinomyces, a gram-positive bacillus, causes infection associated with dental extraction, tooth decay, or oral trauma. Infection begins in the mouth and spreads into the neck,

forming abscess cavities and multiple fistula tracts, which drain thick yellow material. Sulfur granules on microscopic exam are diagnostic. Actinomycosis is treated with penicillin.

Sialadenitis of the submandibular gland may have a viral, bacterial, or inflammatory cause. The gland is tender and warm, with overlying erythema. Pain is often worsened with eating. Patients may describe a foul taste in the mouth from purulent drainage via the salivary duct. Treatment of acute bacterial infection includes antibiotics and maneuvers to decrease salivary stasis: massage, warm compresses, sialagogues, and removal of any obstructing salivary stones. Viral infection is self-limited.

Cat scratch disease is a benign self-limited cause of painful regional lymphadenopathy, fever, and general malaise. *Bartonella henselae* is the causative organism and is commonly transmitted via skin trauma. Patients may give history of a papule 3–5 days after exposure at the site of inoculation. The diagnosis is confirmed with antibody titers and treatment is supportive.

Scrofula is a subacute lymphadenitis caused by *Mycobacterium tuberculosis* or atypical mycobacterium. A thorough exposure history should be obtained if suspected, and a PPD and chest X-ray should be obtained. The most common presentation is subacute unilateral node enlargement and vague systemic symptoms that do not resolve with antibiotic treatment for acute bacterial lymphadenitis. The overlying epidermis becomes discolored, thinned, parchment-like, and adherent to the node. Skin changes may progress to form a sinus tract draining thick white material. A fine-needle aspiration can yield sufficient material for culture but also risks formation of a fistula tract. Treatment is long-term antibiotic therapy. Surgical excision is required for removal of involved skin when skin changes are noted or a fistulous tract has developed.

A neck mass can be the presenting symptom of a patient with *AIDS* (acquired immunodeficiency syndrome). Patients with atypical presentation should receive human immunodeficiency virus (HIV) testing. Patients with known history of AIDS or other immunodeficiency should get skin testing for TB, fungal infections, and excisional biopsy for pathology evaluation for lymphoid hyperplasia, lymphoma, and other malignancies.

Neoplastic Neck Masses

Neoplasms are the most common cause of neck masses in adults but should also be suspected in children with enlarging neck masses that do not resolve in 6–8 weeks and a trial of antibiotics. Malignant neck masses are most commonly *lymph node metastasis* from a primary head or neck malignancy. Initial evaluation is typically with a thorough office exam, CT scan, and fine-needle aspiration (FNA). Excisional biopsy may be necessary for definitive diagnosis of some malignancies such as lymphoma; however, incisional biopsy should not be considered as this is associated with seeding the wound with cancerous cells. Risk factors include tobacco and alcohol use. New studies have shown a close association of squamous cell carcinoma (SCC) of the oral cavity and oropharynx with human papillomavirus (HPV) infection. Patients with HPV-associated oropharyngeal SCC are typically younger, nonsmokers, and nondrinkers and have a better prognosis than HPV-negative tumors.

Lymphatic drainage in the neck follows patterns and directs the search for the primary site. SCC of the jugulodigastric lymph nodes are most commonly sites of metastasis from the oral cavity, oropharynx, and larynx. If no primary source for SCC is evident on office exam, operative panendoscopy (laryngoscopy, bronchoscopy, and esophagoscopy) with directed biopsies and tonsillectomy are performed to locate the primary tumor. A primary tumor escapes detection in up to 20 % of cases.

Other common sources of regional neck metastasis include nasopharyngeal carcinoma and melanoma. A posterior triangle neck mass is the presenting symptom in 60–76 % cases of nasopharyngeal carcinoma. Nasopharyngeal carcinoma is associated with Epstein–Barr virus (EBV) and a high prevalence among people of southern Chinese descent. Isolated left supraclavicular lymph node masses are frequently metastases from chest, abdomen, or pelvic malignancies that drain via the thoracic duct. Management includes chemoradiation, induction chemoradiation and surgical resection, or surgical resection with adjuvant chemoradiation.

Primary malignancies of the neck are uncommonly encountered and are usually *lymphomas*. Enlarged, nontender, rubbery cervical lymph nodes are the presenting complaints of 40 % of patients with Hodgkin's disease. Patients may be asymptomatic or have vague systemic complaints such as low-grade fevers, malaise, or weight loss.

Thyroid masses are most commonly benign nodules. They may be immobile as the thyroid is fixed to the trachea via the ligament of Berry, and the gland elevates with swallowing. A diffusely enlarged thyroid is typically multinodular goiter. A dominate nodule should be evaluated by ultrasound and ultrasound-guided FNA. Risk factors for thyroid cancer include multiple endocrine neoplasia type 2 (MEN-2) syndrome and history of radiation exposure.

Benign neoplasms of the neck are uncommon. A carotid body tumor (paraganglioma) or vagal schwannoma presents as a lateral anterior triangle neck mass. Carotid sheath tumors are firm and move side to side but not up and down. Pulsatile masses or masses with a bruit are indicative of a vascular lesion and FNA should be avoided. Initial evaluation is with CT scan. Paragangliomas are usually benign (rate of malignancy around 10 %), but their high vascularity and location make resection difficult.

Part VII
Pediatrics

70
Hypertrophic Pyloric Stenosis

Michael W. Dingeldein

Presentation

Nonbilious projectile emesis between weeks of life 2–5 with hypochloremic, hypokalemic metabolic alkalosis is the cardinal feature of hypertrophic pyloric stenosis (HPS) (Fig. 70.1). The vomiting often happens soon after feeding, with the child continuing to act hungry and appearing vigorous without fevers. The health of the child at presentation can vary widely due to hydration status, from well appearing to lethargic and severely dehydrated. Not uncommonly, children present with HPS relatively late in the disease course after changes in feedings or formulas have been made with the thought that the emesis was secondary to feed intolerance or reflux.

Etiology/Pathophysiology

HPS is the most common cause of gastric outlet obstruction in infants. It was first described on a postmortem exam in 1717 by Blair. Hirschsprung published the first complete clinical description. Ramstedt in 1912 was the first to do the standard modern operation of muscle splitting.

The cause of the gastric outlet obstruction is exactly as the name implies: the pyloric muscle is hypertrophic causing an occlusion. Why this happens is not as clear. There is a genetic component with increased incidence in *first-born males with a positive family history*. Multiple cytokines and growth factors such as substance P, gastrin, EGF, neurotrophins, and somatostatin have been noted to be increased in HPS, but none have been shown to have a direct relationship. Alterations in the metabolism of nitric oxide within the pylorus have been of interest in recent research, but again direct relationships have proven elusive.

Evaluation

Evaluation is straightforward, and often the diagnosis of HPS can be made with a good history and physical exam alone. It is important to rule out other causes of infant emesis such as gastroenteritis, food allergy, and reflux. Physical exam is focused on hydration status and finding an "olive," which is an enlarged pylorus in the upper abdomen just underneath the liver edge in the midline. Fevers and leukocytosis are rare in HPS.

Abdominal ultrasound has become the standard imaging modality for HPS. Single pyloric wall thickness greater than 4 mm and pyloric channel length greater than 14 mm are generally accepted criteria. In addition, there should not be any gastric contents seen passing through the pyloric channel.

Measurement of basic electrolytes such as sodium, potassium, chloride, and bicarb is mandatory.

Treatment

There is no such thing as an emergent pyloromyotomy. Rehydration and correction of electrolyte abnormalities are always the first treatment priority in HPS. Aggressive intravenous resuscitation with an isotonic dextrose containing crystalloid fluid is key. Basic goals include urine output of

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_70,

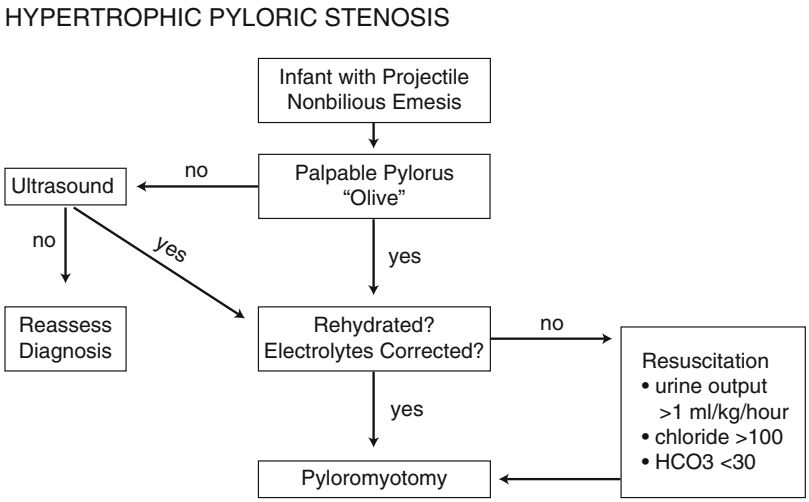

FIG. 70.1 Treatment algorithm for hypertrophic pyloric stenosis. *HCO3* bicarbonate

greater than 1 ml/kg/h and chloride greater than 100 with serum bicarb less than 30.

After appropriate resuscitation is accomplished, a pyloromyotomy is done based on Ramstedt's basic principle that you can divide the pylorus muscle without having to repair it as long as the mucosa is not violated.

Surgical treatment is the same regardless whether it is done open or laparoscopically. The pylorus muscle is bluntly divided longitudinally after scoring the serosa. The clean-appearing mucosa should bulge between the divided sides of the muscle. If the mucosa is entered, it can be repaired with fine absorbable suture and buttressed with omentum. Alternatively, the seromuscular layer can be closed overtop the open mucosa and a new pyloromyotomy site can be chosen.

Postoperative Management

It is common for a child with HPS to have some episodes of emesis post-op. Feeding is started 6 h post-op with low volume electrolyte solution (pedialyte) and advanced in volume slowly every 3 h. After the first few feedings, it can then be switched to formula or breast milk. Once intravenous fluids have been stopped and the infant is tolerating full feeds, the child can be discharged – usually 1–3 days post-op.

Long-term prognosis is excellent. Inability to tolerate feeds postoperatively is not due to recurrent HPS but usually due to insufficient division of the pylorus.

71
Jaundice in the Pediatric Patient

Srikumar Pillai

Introduction

Clinically apparent jaundice in a neonate generally occurs when the serum bilirubin exceeds 5 ml/dl. This level may be somewhat lower for an older child. The bilirubin can be either unconjugated (indirect) or conjugated (direct), with the differential diagnosis for each varying significantly (Fig. 71.1).

Physiologic jaundice (an unconjugated hyperbilirubinemia) is the most common cause of jaundice in the newborn. It occurs in approximately 15 % of full-term newborns in the first 2 weeks of life. It has a multifactorial etiology, primarily due to immaturity of the enzyme glucuronyl transferase. Immaturity of this enzyme, which is responsible for bilirubin conjugation, leaves neonates more prone to cholestasis and therefore more prone to jaundice. Serum bilirubin concentration in full-term newborns should be less than 13 mg/dL and usually resolves within a week. Other causes of unconjugated hyperbilirubinemia include breast-milk jaundice, hemolytic disorders, hypothyroidism, and familial disorders. Surgically treatable disorders include pyloric stenosis and other forms of intestinal obstruction.

Direct Hyperbilirubinemia

Direct hyperbilirubinemia is considered pathologic when the fraction of conjugated bilirubin is greater than 20 % of the total serum bilirubin concentration (when the total bilirubin is greater than 5 mg/dL) or when the conjugated bilirubin level is greater than 1 mg/dL. Jaundice of this degree that persists longer than 2 weeks of life should be evaluated immediately. Direct hyperbilirubinemia results from either hepatocellular disease where conjugated bilirubin cannot be excreted out of the hepatocyte into the bile duct caniculi or from functional or mechanical/obstructive cholestasis.

The causes of direct hyperbilirubinemia vary by age. Therefore, different diagnoses should be considered when looking at the neonate as compared to the older child. Surgical conditions in older children include cholelithiasis/cholecystitis and choledochal cyst. However, many medical conditions may progress to jaundice and end-stage liver disease requiring transplantation, and these should be considered.

Direct hyperbilirubinemia in an infant requires prompt diagnosis because all of the possible etiologies are serious and need urgent treatment. Medical causes include sepsis due to perinatal infections, such as TORCH infections (*t*oxoplasmosis, *o*ther [syphilis], *r*ubella, *c*ytomegalovirus, *h*erpes simplex virus), or urinary tract infections (UTIs), primarily *Escherichia coli*. Metabolic and inherited conditions such as α(alpha)-1-antitrypsin, cystic fibrosis, and tyrosinemia must also be considered. These are diagnosed with various blood and urine tests and cultures. Surgical causes include anomalies of the liver and biliary tree, principally biliary atresia, and choledochal cyst. The three most common causes of cholestasis in the neonate are idiopathic neonatal hepatitis, biliary atresia, and α(alpha)-1-antitrypsin deficiency.

The workup of these conditions must proceed in a timely fashion. While cultures and serologic results are pending, evaluation for possible biliary atresia can be initiated with an abdominal ultrasound. This will help identify any choledochal cyst or other common duct abnormalities and will help determine whether a gallbladder is present. If a choledochal cyst is identified, no other workup is necessary. Absence of the gallbladder is suggestive of biliary atresia; although, its presence does not exclude the diagnosis since the gallbladder may not be in circuit with a patent biliary tree but instead may be filled with mucus or "white bile."

A hepatobiliary iminodiacetic acid (HIDA) scan can be useful in distinguishing between obstructive and parenchymal causes for direct hyperbilirubinemia. Quick hepatocyte uptake without excretion into the gastrointestinal tract suggests biliary atresia, while poor uptake is indicative of hepatocyte dysfunction as seen in idiopathic neonatal hepatitis. Poor hepatocyte uptake, and therefore HIDA scan accuracy, may be improved with pretreatment with phenobarbital, which increases biliary flow. A percutaneous liver biopsy may help in making the distinction between atresia and hepatitis by

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_71, © Springer New York 2015

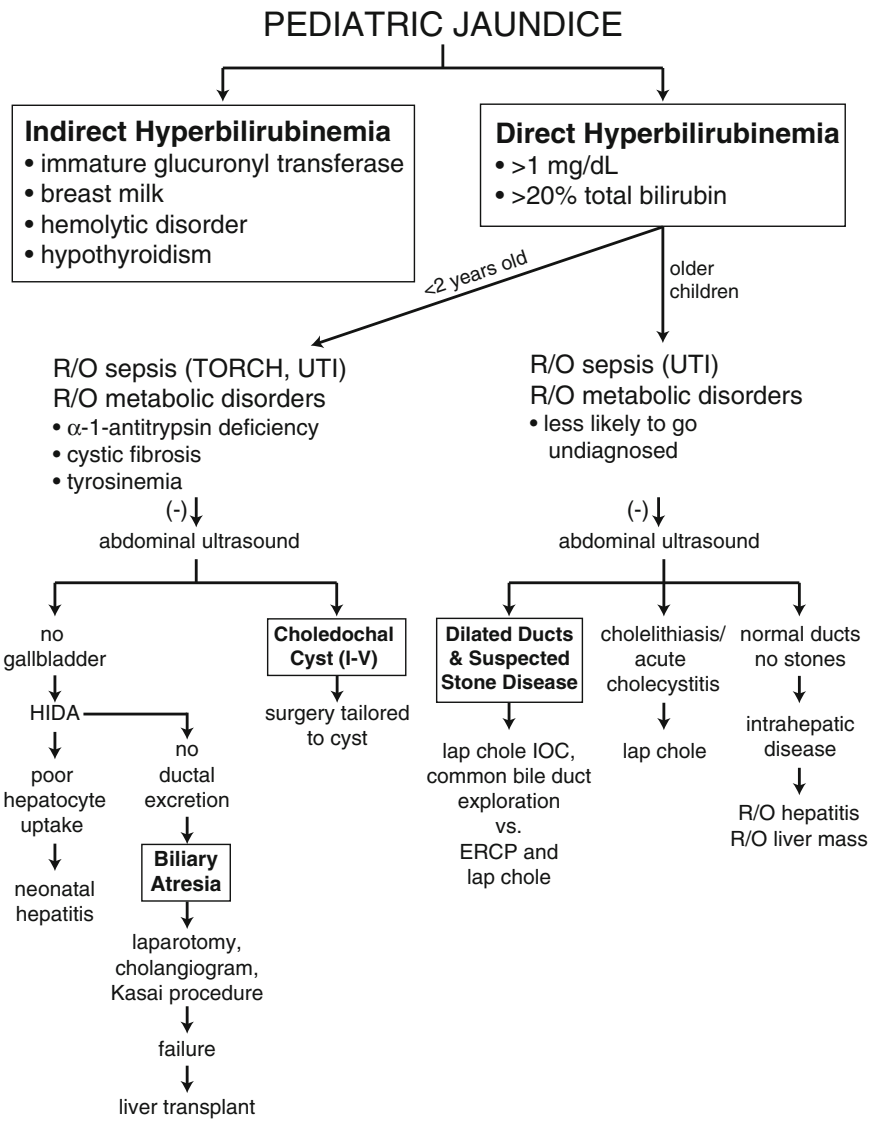

FIG. 71.1 Treatment algorithm for jaundice in pediatric patients. *R/O* rule out, *TORCH* toxoplasmosis, other (such as syphilis, varicella, mumps, parvovirus, HIV), rubella, cytomegalovirus, herpes simplex; *UTI* urinary tract infection; *HIDA* hepatobiliary iminodiacetic acid; *lap chole IOC* laparoscopic cholecystectomy with intraoperative cholangiogram; *ERCP* endoscopic retrograde cholangiopancreatography

revealing either bile duct proliferation as seen with biliary atresia or focal necrosis seen with hepatitis. However, there is some overlap of these histologies. The diagnosis is firmly established by laparotomy and intraoperative cholangiogram, demonstrating either the absence of a functional gallbladder or the lack of biliary continuity between the liver and the duodenum.

Biliary Atresia

Biliary atresia is characterized by progressive obliteration of the extrahepatic biliary tree, proliferation of small intrahepatic bile ducts, the presence of plugs of inspissated bile within the caniculi, and periportal fibrosis. The etiology for the pathogenesis of biliary atresia has not been clearly established. These patients present in the first few weeks of life with jaundice and hepatomegaly but generally appear well in contrast to patients with an infectious etiology. Initially, stools may have normal color but soon become acholic, consistent with the progressive nature of biliary atresia. The only treatment available is surgical treatment, and without it patients will develop biliary cirrhosis and portal hypertension and will die within 2 years.

A Kasai hepatoportoenterostomy is performed for biliary atresia by carefully dissecting out the distal bile duct towards the liver capsule. It is then excised along with any gallbladder remnant that is present. Finally, an anastomosis is made between the small bowel and the exposed liver parenchyma. Despite a technically well-performed operation, the results from this procedure are variable. About one-third of patients will have a good result and remain anicteric. Another third will never drain bile and progress to liver failure. The remaining patients often get some relief from their jaundice, but over a long period of time ultimately suffer end-stage liver failure. One factor in the prediction of

outcome for these patients is the age at operation. Those performed within the first 2 months of life have a significant survival advantage, whereas there is probably no likelihood of success after 4 months of age. The only salvage for patients with a failed Kasai is liver transplantation.

Choledochal Cyst

Choledochal cyst is a congenital enlargement of the biliary tree and can be categorized into five subtypes according to modification of the Alonso-Lej classification:

- I – cystic dilation of the common bile duct (80–90 % of cases)
- II – diverticulum of the common bile duct
- III – choledochocele, found in the intrapancreatic portion of the bile duct and often involves the ampulla
- IV – intrahepatic and extrahepatic dilations (second most common)
- V – intrahepatic cysts only (Caroli's disease)

Choledochal cysts should be considered when a patient presents with abdominal pain, right upper quadrant mass, and jaundice; 85 % of pediatric patients with a choledochal cyst will present with two out of these three symptoms. Infants with choledochal cysts will often present with jaundice as their only symptom, while older children tend to have abdominal pain and abdominal mass. Pancreatitis can also be seen on presentation. This condition is more common in females and Asians. Its pathogenesis is not understood. It is theorized that this condition may be a result of pancreatobiliary reflux or possibly due to obstruction of the distal bile duct.

The diagnostic modality of choice is an abdominal ultrasound. If dilation of the extrahepatic bile ducts is seen, no additional testing is necessary. If uncertainty remains, a hepatobiliary radionuclide scan or computed tomography (CT) may provide additional information. In older patients, an endoscopic retrograde cholangiopancreatography (ERCP) can be done to confirm diagnosis. The treatment for Type I cysts is cyst resection with internal biliary drainage established via Roux-en-Y hepaticojejunostomy. Cholecystectomy is also usually performed. If the cyst is densely adherent to the portal vein, the posterior cyst wall may be left in place after the inner lining of the cyst wall has been excised. Simple cyst-enterostomy should be avoided because of the high incidence of stricture formation and the possibility of future malignancy. Type II cyst may be treated in the same fashion as the Type I cyst or with diverticulectomy. Type III requires transduodenal cyst unroofing and sphincteroplasty. Type IV and V are more challenging and may require segmental liver resection or even liver transplantation.

Stone Disease

Biliary obstruction secondary to stone disease should be considered in older children presenting with jaundice. Stones may become lodged in the common bile duct, just as in adults with choledocholithiasis, causing an obstructive hyperbilirubinemia. These patients may present with jaundice, right upper quadrant or epigastric pain, lightening of stools, and darkened urine. The addition of fevers and mental status changes should alert the physician to the possibility of cholangitis.

In working these patients up, lab results may reveal a hyperbilirubinemia as well as elevations in alkaline phosphatase and the transaminases. Ultrasound is a very useful modality in evaluating biliary disease. It is helpful in identifying cholelithiasis/acute cholecystitis as well as dilation in the common bile duct indicating possible choledocholithiasis. Cholecystitis and cholelithiasis are treated surgically with a cholecystectomy. If choledocholithiasis is suspected, surgical treatment involves a laparoscopic cholecystectomy with intraoperative cholangiogram and common bile duct exploration. Alternatively, endoscopic retrograde cholangiopancreatography (ERCP) may be done preoperatively for stone extraction followed by a laparoscopic cholecystectomy. In older children, if on ultrasound the biliary ducts appear normal and there are no signs of stones, hepatitis and possible liver mass should be considered.

72
Pediatric Abdominal Masses

Michael W. Dingeldein

General Considerations

The differential diagnosis for pediatric abdominal masses is extensive but can be quickly narrowed down by doing a complete history and physical exam and considering the patient's age and symptoms (Fig. 72.1). Basic imaging such as plain abdominal X-rays and ultrasound are very helpful. Advanced imaging with computed tomography (CT) and magnetic resonance imaging (MRI) often establishes the diagnosis.

Congenital Anomalies

Gastrointestinal (GI) Tract

There are a variety of congenital GI problems that cause abdominal distention. Duplications of the GI tract can occur anywhere. There is a 1:4,500 incidence, with the jejunum and ileum being the most common locations. Many of these lesions are closed loops of bowel and over time will distend with fluid, giving the appearance of a large cystic mass. Surgical resection is the treatment of choice.

Mesenteric and omental cysts are rare, representing 1:20,000 pediatric hospital admissions. They are commonly thought to be caused by failure of the lymphatic system to properly form. Surgical resection is the treatment of choice if they are symptomatic. Omental cysts are relatively easy to remove. Mesenteric cysts can be complicated to excise and doing so can compromise the intestinal blood supply. Lymph leakage is another common complication that may occur after surgery.

Genitourinary (GU) Tract

There are an assortment of GU conditions that present as an abdominal mass. Bladder obstruction can present as a large anterior abdominal mass and may be asymptomatic, especially in nonverbal children. Ureteral duplication and megaureter are rare malformations that can appear as large fluid-filled masses. Pediatric renal masses are overall rare but are on the list of pediatric abdominal malignancies (see Wilms tumor below). Polycystic kidney disease is seen primarily in teenagers and can be easily diagnosed by ultrasound.

Congenital vaginal obstruction due to imperforate hymen, agenesis of the lower vagina, or transverse vaginal septums can present in a variety of ways in which abdominal distention may accompany hydrocolpos, hydrometrocolpos, mucometrocolpos, or pyometrocolpos.

Ovarian masses are relatively common and discussed below.

Tumors

Only 2 % of all cancer cases occur in childhood, but after trauma it is the number 2 cause of death in children. Twelve thousand children under the age of 18 develop cancer in the USA per year, most being leukemia, brain tumors, and lymphoma, but a significant number of solid tumors, such as neuroblastoma and Wilms, present as abdominal masses.

Neuroblastoma

Neuroblastoma is the most common solid tumor of childhood, representing 10 % of all childhood tumors and 15 % of childhood cancer deaths. It originates from neuroblasts in the sympathetic ganglion chain anywhere along its course from the neck to pelvis. The evolution of the disease and its behavior varies as widely as its anatomic location, from spontaneous regression to severely progressive disease. Its presentation varies depending on tumor location, but in 75 % of cases it is as an abdominal mass and has generalized symptoms of weight loss, abdominal pain, and failure to thrive. Twenty-five percent will have hypertension or other signs of increased

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_72, © Springer New York 2015

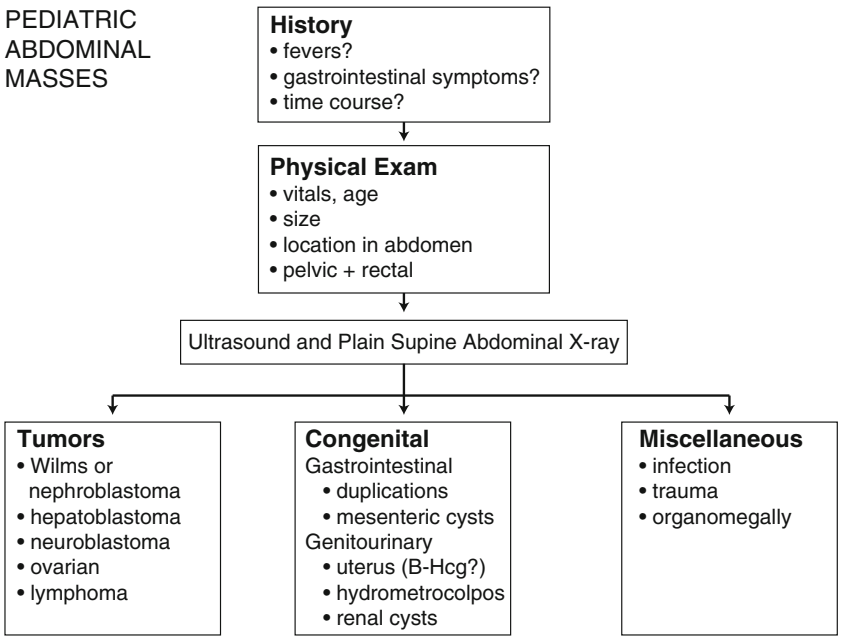

FIG. 72.1 Algorithm for treatment of pediatric abdominal masses

catecholamine release (flushing, sweating, irritability). Metastases to the bony orbit produce the classical presentation of "raccoon eyes." Treatment is multidisciplinary with complete surgical resection being the cornerstone and augmented by chemotherapy and radiation.

Wilms Tumor

Wilms tumor (aka nephroblastoma) is the most common renal tumor of childhood (91 %) and represents about 6 % of all childhood tumors. In the USA, roughly 500 cases are diagnosed per year. There have been striking improvements in outcomes over the last 50 years. Long-term survival in developed countries now exceeds 85 % overall and 99 % for low-stage tumors. Most children with Wilms tumor present with an asymptomatic abdominal mass; 20 % have gross hematuria and 25 % have hypertension from renin-angiotensin activation. A left varicocele is a rare presentation caused by tumor extension into the renal vein. Diagnosis is made by physical exam and CT scan. Most common metastatic spread is to the lungs and liver. Screening exams are recommended for Beckwith-Wideman syndrome, Perlman syndrome, and idiopathic hemihypertrophy. The two most important prognostic factors are histology and stage. Surgical resection is the treatment of choice, especially for low-stage tumors. Some centers advocate preoperative chemotherapy, particularly in high-risk tumors.

Liver Tumors

Primary liver tumors constitute about 3 % of all pediatric tumors and can be either benign or malignant. Most present as large RUQ masses with vague abdominal symptoms such as dull pain, early satiety, and vomiting. It is not uncommon to have jaundice and weight loss. Workup consists of serum alpha fetoprotein (AFP), abdominal ultrasound, and/or a triple phase CT scan of the liver. The most common *benign liver tumor* is a hepatic hemangioma, which is classified as focal, multiple, or diffuse. These are not malignant lesions and treatment is surgical. Prognosis for focal and low-volume multiple hemangiomas is excellent. The diffuse form has a survival of about 75 %. Hepatic adenomas and focal nodular hyperplasia occur in children but are rare compared to adults. They are treated in the same fashion as adults.

There are two primary types of *malignant pediatric liver lesions:* hepatoblastoma (HB) and hepatocellular carcinoma (HCC). In the USA, 80 % are HB, but in developing countries there is a much higher incidence of HCC. HB is actually a collection of a variety of histologies all based on distinct phases of hepatogenesis. Treatment is complete surgical resection followed by chemotherapy. For large or multifocal tumors, "down staging" with preoperative chemotherapy has been tried. Transplant is an option for some tumors. HCC is associated with older children and teenagers (60 % over the age of ten), particularly those that are hepatitis B positive. It is also associated with alpha-1 antitrypsin deficiency, alagille syndrome, biliary atresia, and chronic TPN. HCC is poorly responsive to chemotherapy, and the best hope for cure is complete surgical resection.

Ovarian Tumors

The majority of ovarian masses in children are not malignant (most occur in girls aged 15–19 years). The most common

neoplasms are germ cell tumors, followed by epithelial tumors, stromal tumors, and then miscellaneous tumors. Non-germ cell tumors include epithelial ovarian neoplasia and sex cord-stromal tumors. Both are rare and treated by resection.

Presentation is most often with vague symptoms of abdominal pain, but ovarian torsion is a classic presentation with high morbidity. There is a 1 % rupture rate for all ovarian masses. Treatment is primarily surgical resection, and cure rates approach 100 % in low-stage tumors.

The most common germ cell tumor of the ovary is a teratoma. Teratomas are divided into two categories: mature and immature. Mature teratomas tend to have more developed tissues, where as immature are more homogenous. Mature cystic teratomas are almost all benign but have a .2 % to 2 % rate of malignant transformation. Risk of malignant disease is higher with immature ovarian teratomas. Low-stage tumors have an excellent prognosis, but advanced tumors can be aggressive. The mainstay of treatment for ovarian teratomas is resection.

73
Esophageal Atresia/Tracheoesophageal Fistula

Kristin Bevil

Incidence and Variants

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) are congenital abnormalities that occur in approximately 1 in 4,000 births (Fig. 73.1). These abnormalities are related to defective orientation of the tracheoesophageal septum as the trachea forms from the caudal foregut during the 6th week of gestation. There are five basic variants. EA with normal tracheal development is designated as type A. Unlike the other types, it is due to a defect in recanalization of the esophagus in the 8th week of gestation, and it occurs in 80 % of cases. Type B is EA with proximal TEF, occurring in 1 % of cases. *The most common variant (85 %) is type C, consisting of EA with distal TEF*. The TEF is typically located near the level of the carina. EA with both distal and proximal TEF, type D, occurs in 2 % of cases. Finally, type E consists of TEF with no EA. The defect is usually located high on the trachea and occurs in 4 % of cases. It is also known as H-type TEF.

Associated Defects

Approximately half of neonates with EA/TEF have additional malformations. These defects most commonly fall into the **VACTERL** association, composed of three or more of the following abnormalities: vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb. Of infants with TEF, 23 % have cardiac, 18 % have musculoskeletal, 16 % anorectal or intestinal, and 15 % have genitourinary abnormalities. A high suspicion for these associated defects enables more rapid diagnosis and initiation of appropriate management.

Presentation

EA/TEF may be suspected during prenatal surveillance with **polyhydramnios** on ultrasound secondary to defective swallowing. Infants present early in the neonatal period with **feeding intolerance** and varying levels of respiratory distress. Despite suctioning, infants have copious frothy white mucus secretions from the mouth and, occasionally, the nose. In addition, they may have episodes of **coughing and choking** with possible **cyanosis**. These cyanotic episodes may be worse with feeding, especially with a proximal TEF. A distal TEF may lead to tympanitic abdominal distension and aspiration of gastric contents. H-type TEF may present later in life with more subtle signs of aspiration, such as recurrent pneumonia. Physical findings of VACTERL abnormalities may also be immediately visible, such as an imperforate anus or limb malformations.

Diagnosis

Initial evaluation of a suspected EA is done with attempted passage of a radiopaque nasogastric tube. The tube typically fails to pass more than 10–12 cm (the stomach is approximately 17 cm from the mouth). X-ray of the baby shows the curled tube in the neck or upper chest, confirming the diagnosis. X-ray may also show intraluminal gas under the diaphragm, indicating a distal fistula (type C or D), while a lack of gas indicates the absence of or a proximal TEF (type A or B). The X-ray also aids in identification of other VACTERL abnormalities. Bronchoscopy is required to diagnose an H-type TEF as a plain film is typically normal. It also helps determine the precise level of any TEF.

Medical Management

Once diagnosed, immediate management revolves around aspiration precautions and transfer to a tertiary care center with a neonatal intensive care unit. Aspiration precautions include continuous suctioning of the proximal pouch, elevation of the infant's head, fluid resuscitation, and total parental nutrition (TPN) until surgery is performed. Oxygen is administered as needed; endotracheal intubation with a cuffed tube is performed if respiratory failure develops. Bag-mask ventilation is avoided in order to prevent gastric

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_73, © Springer New York 2015

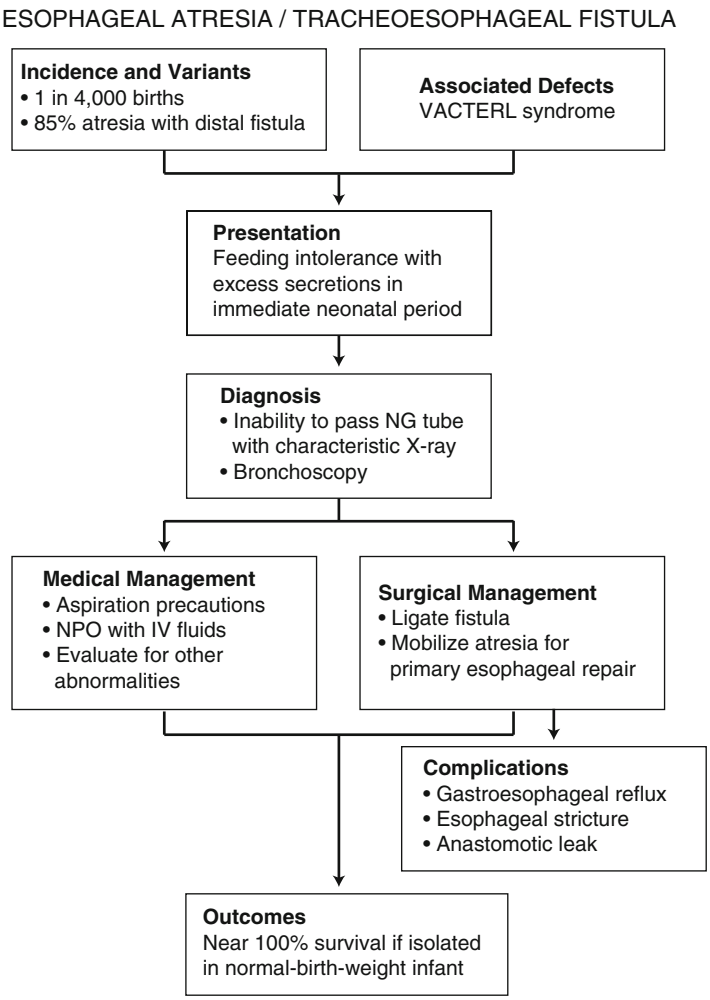

FIG. 73.1 Treatment algorithm for esophageal atresia/tracheoesophageal fistula. *VACTERL* vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb; *NG* nasogastric; *NPO* nil per os; *IV* intravenous

distension. Broad-spectrum antibiotics are used in infants with suspected sepsis or pneumonia. Evaluation for other VACTERL abnormalities occurs during this presurgery time as well, consisting of chest and abdominal X-ray, echocardiogram, and renal ultrasound.

Surgical Management

Surgery can often be performed within the first few days of life. Surgical correction requires ligation of the TEF and primary repair via esophagoesophagostomy. Bronchoscopy may be performed in the operating room if not performed previously in order to determine the level of the TEF, look for an additional TEF (usually high H-type), and aid in placement of the cuffed endotracheal tube. The defect is approached from a right lateral thoracotomy incision. The TEF is identified behind the azygos vein and ligated. The trachea may be closed with a flap of mediastinal pleura to avoid repeat fistula formation. The proximal esophageal pouch is then identified and mobilized, if needed. The distal esophagus may be mobilized, but care should be taken to protect and preserve its blood supply. Once the ends are approximated, a single-layer, full-thickness anastomosis is performed with interrupted sutures. A chest tube is placed to drain potential anastomotic leaks. The incision is closed with tension-free approximation of the ribs to avoid fusion. Esophagram may be performed 5–7 days post-surgery, and, if no leak is detected, oral feedings may be started.

Special Circumstances

Surgery should be delayed in infants with low birth weight, pneumonia, sepsis, or other significant abnormalities. Major cardiac abnormalities should be corrected prior to surgery. Alternatively, surgery may be performed in stages with initial ligation of the TEF followed by EA repair in infants who are premature or in severe respiratory distress.

Long-gap EA (usually type A) is defined as a distance of >3 cm or >2 vertebral bodies. This variant provides a unique surgical challenge as tension on the anastomosis can lead to significant stricture and anastomotic leaks. These patients are best treated by initial gastrostomy placement, ligation of the TEF if present, followed by watchful waiting in case the pouches grow closer as the infant grows. Esophageal dilatations

may promote esophageal growth and may be performed via pressure on the nasogastric tube in the proximal pouch or magnets or external sutures. After 3 months of this, patients undergo repair via esophagoesophagostomy. If the gap is still too wide, esophageal reconstruction can be done via gastric pull up or colonic or jejunal interposition.

H-type defects (type E) may be accessed via left cervical incision due to their high placement on the trachea. The fistula is identified and divided. A stent or catheter placed with the help of bronchoscopy prior to surgery may aid in identification.

Complications

Anastomotic leak is an early surgical complication that occurs in 15–20 % of patients, typically as a result of tension on the anastomosis or ischemia due to mobilization. Most resolve spontaneously within a few days. Major leaks require repeat thoracotomy with esophageal replacement. The most common reason for repeat surgery, however, is esophageal stricture. Stricture occurs in 30–40 % of cases and may be responsive to esophageal dilatation. Gastroesophageal reflux occurs in 40–65 % of patients and may be managed medically or surgically. *Nearly all patients have esophageal dysmotility*.

Outcomes

Isolated EA/TEF in normal birth weight infants is associated with near 100 % survival. Worse prognosis is associated with very low birth weight prematurity, major associated abnormalities, and long-gap EA.

74
Intussusception

Jarod P. McAteer, Morgan K. Richards, and Adam B. Goldin

Introduction

Intussusception is a common cause of bowel obstruction in infants and young children. The condition occurs when the intussusceptum is invaginated into the intussuscipiens. In more than 80 % of cases, the distal ileum invaginates into the colon (termed ileocolic intussusception). Although most cases are idiopathic, a pathologic lead point may be identified in 2–12 % of children, the likelihood increasing with age. After age 4 years, lead points are found in over 50 % of patients. In adult cases of intussusception, an identifiable lead point is present in as many as 97 % of cases. Lead points are generally intraluminal or intramural anatomic abnormalities. In children, the most common lead point is Meckel's diverticulum, followed by ileal and colonic polyps, intestinal hamartomas, submucosal hemorrhages associated with Henoch-Schonlein purpura, lymphoma, lymphosarcoma, enteric cysts, inverted appendiceal stumps, and anastomotic suture lines. In idiopathic cases, the most common pathologic finding is hypertrophy of Peyer's patches in the ileal wall, which is common during and immediately following viral infections of the respiratory and gastrointestinal tracts. Adenoviruses have frequently been implicated in the etiology of Peyer's patch hypertrophy. Regardless of etiology, as intussusception evolves, the mesentery of the proximal bowel segment is drawn further into the distal segment, resulting in venous obstruction and eventual arterial insufficiency and bowel necrosis if the obstruction is not relieved.

Surgeons evaluating children under the age of 5 with acute abdominal pain should have a high suspicion for intussusception, which causes up to 25 % of abdominal surgical emergencies in this age group (Fig. 74.1). The peak incidence in infants is between 5 and 9 months of age, and 80–90 % of all cases occur in children ages 3 months to 3 years. Most patients are otherwise healthy children, and up to two-third of cases occur in males. The condition commonly presents as intermittent, crampy abdominal pain beginning suddenly in a previously well-appearing child. The pain is often described as colicky and short in duration, and returns in 20–30 min intervals. Emesis is common and may follow the pain attacks, which are often accompanied by writhing, breath holding, and fetal position posturing. The emesis consists initially of undigested gastric contents, later progressing to bilious content as the obstruction progresses. Bowel movements also may accompany pain attacks and are usually normal early in the disease course. As ischemia progresses, the bowel lining may slough, resulting in "currant-jelly" stools. *The "classic" triad of intermittent pain, emesis, and currant-jelly stools is seen in less than 20 % of cases*. While patients may initially appear well with unimpressive symptoms between attacks, lethargy becomes common as the disease progresses, and delay in diagnosis can lead to bowel ischemia, perforation, sepsis, and shock. Cardiovascular collapse and death are possible if prompt treatment is not initiated.

Diagnostic Evaluation

Early in the disease, the patient's vital signs may be normal, but tachycardia becomes prominent as the disease progresses. Between pain episodes, a flat, empty right lower quadrant may be noted on physical exam, as the cecal portion of the intussusception is carried toward the hepatic flexure and transverse colon. The classic "sausage-like" mass may be found anywhere in the abdomen, although its presence is noted in only 15 % of cases. These findings can be very difficult to elicit when the patient is in pain and the abdomen is tense. On rectal exam, many children will have gross blood, mucoid bloody stools, or hemoccult positivity. Rarely, the examiner may note the intussusceptum prolapsed through the anus. In such cases, it is essential to differentiate prolapsed intussusception from simple rectal prolapse.

The key to definitive diagnosis in most cases is imaging. Abdominal radiographs are frequently used but are largely nonspecific. Findings are normal in about 25 % of patients. Signs of small bowel obstruction may be seen in up to half of patients, but are not reliable unless the illness has been present for some time. The classic finding of a paucity of right

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_74, © Springer New York 2015

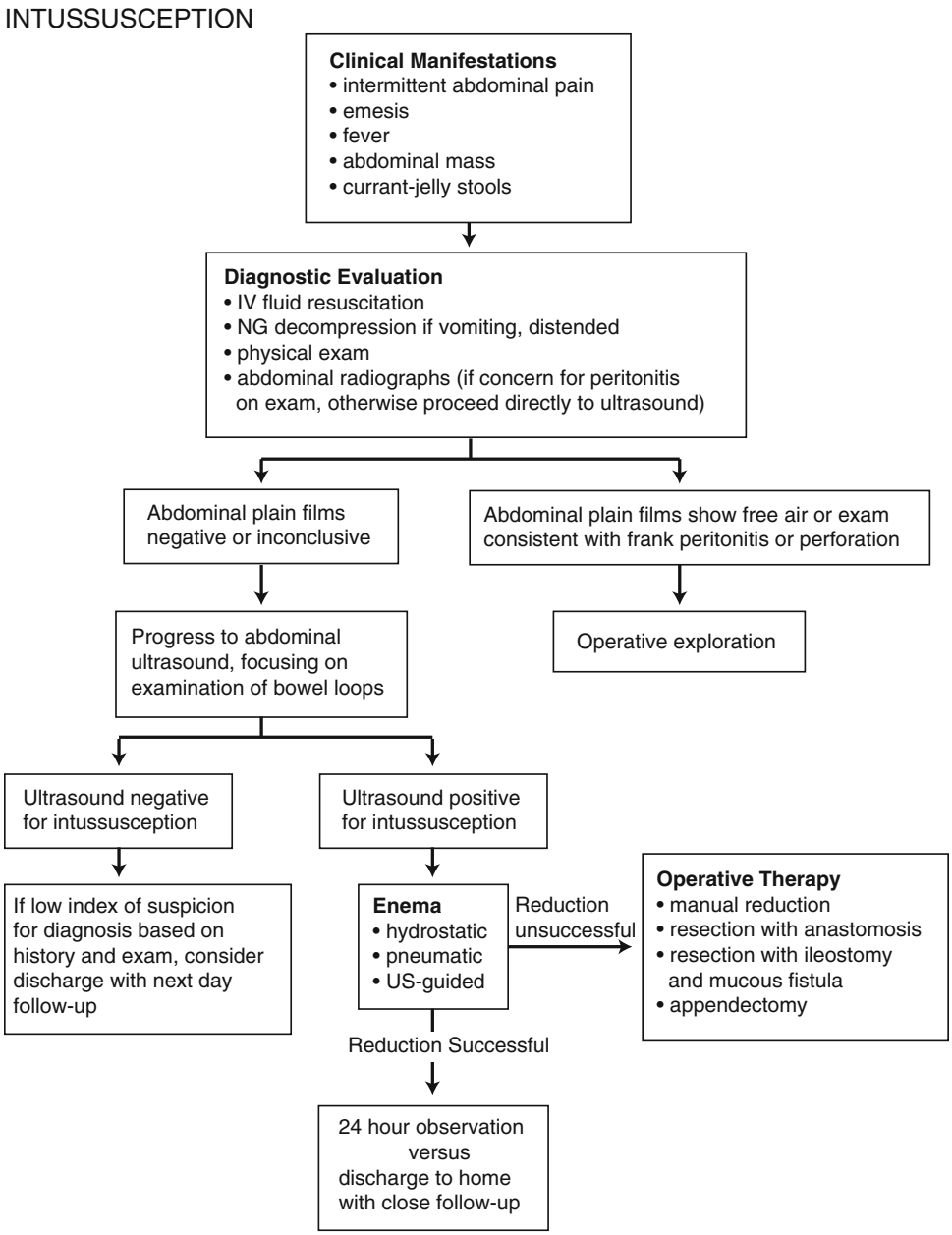

Fig. 74.1 Intussusception treatment algorithm. *IV* intravenous, *NG* nasogastric, *US* ultrasound

lower quadrant gas is noted in only 10 % of cases. Because of the limitation of plain films, *abdominal ultrasound is regarded as a more reliable diagnostic modality* (97.9 % sensitive, 97.8 % specific). Characteristic sonographic findings include the **"target" lesion**, consisting of two hypoechoic rings separated by a hyperechoic layer.

Nonoperative Therapy

Initial management should consist of nasogastric tube placement and intravenous fluid administration. Antibiotics with enteric coverage should generally be administered prior to an attempt at reduction. A surgeon should be involved early in the management of these patients, as signs of peritonitis, perforation, sepsis, or bowel necrosis should prompt immediate operative intervention. *In the clinically stable child, nonoperative reduction* via *enema is considered first-line therapy*. There are three basic methods to achieve this.

Hydrostatic reduction involves placement of a noninflatable rectal tube and **administration of barium or another contrast material by gravity**. The buttocks are taped firmly together to hold the lubricated tube in place, and the child is restrained. The height of the fluid column should be adjusted to create an *intraluminal pressure of no greater than 120 mmHg*. The influx of contrast is monitored under

fluoroscopy, and the procedure is continued as long as there is clear progress in reducing the intussusception. If progress ceases, the contrast material should be allowed to drain, although the reduction can then be attempted again the second or third time. Successful reduction should be confirmed by reflux of contrast into the terminal ileum, though, on occasion, edema of this region may hinder this finding. Considering this possibility, some suggest close observation rather than operative therapy if reduction is believed to have been successful, but contrast reflux into the terminal ileum is not observed.

A second method involves **pneumatic reduction under fluoroscopic guidance**, which has become the preferred method of reduction due to higher success rates compared to contrast enemas in large published series. Air is insufflated into the colon and the intussusception is visualized and progress of reduction is constantly monitored. Maximum safe pressures are 80 mmHg in young infants and up to 120 mmHg in older children. It is believed by some that this method is safer as it allows constant monitoring of intraluminal pressure, as well as creates a more evenly distributed pressure along the length of the colon. Drawbacks compared to contrast enema include poor visualization of lead points and the possibility of tension pneumoperitoneum in cases of perforation. Although rates of perforation in the literature are 1 in 250–300 cases, it is advisable that reductions only be attempted with an available surgeon and angiocatheters at the bedside for emergent decompression of tension pneumoperitoneum.

A third method of radiographic reduction relies on **ultrasound-guided hydrostatic** or pneumatic reduction. Hydrostatic methods rely on saline and water-soluble contrast solution so that reflux of contrast into the terminal ileum can confirm successful reduction with an abdominal plain film. Ultrasound can be used to confirm the resolution of the intussusception. Sonographically guided methods are infrequently used, as they rely on a talented sonographer, and image quality can be impaired by bowel gas in cases of significant bowel obstruction.

If an initial trial of enema reduction is not successful, it is reasonable to try again within a few hours as long as the patient's abdominal exam has not markedly worsened. If progress is not made on repeat attempts after 3–5 min of maximum pressure, then further attempts at enema reduction should be abandoned.

If enema reduction is successful, traditional management includes admission and observation with NPO status for a period of 24 h to monitor for recurrence. Recent data suggests that because early recurrence (<24 h) is relatively infrequent, selected patients may be safely discharged to home from the Emergency Department following successful nonoperative reduction. Although post-reduction antibiotics are often used, there is little evidence to support this practice.

Operative Therapy

Children who have clinical or radiographic evidence of peritonitis or perforation, and children who fail radiologic reduction, should undergo operative reduction. A residual filling defect on contrast enema following successful radiographic reduction is also a relative indication for elective surgical management, as this finding generally represents a pathologic lead point. Preoperatively, patients should receive nasogastric decompression, resuscitation with intravenous fluids, and appropriate prophylactic antibiotics. The operation is typically begun with a transverse right lower quadrant muscle-splitting incision. Laparoscopic exploration is also an option but should be converted to an open approach if initial attempts at reduction are not successful. The intussusception is delivered into the wound, and reduction should proceed by gentle compression of the distal end, rather than pulling of the proximal end. If resistance to reduction results in tearing of the bowel, resection of the intussuscepted segment should be considered in order to avoid perforation and spillage. If vascular compromise is suspected, the surgeon should observe the bowel for 10–15 min prior to performing a resection. Warm saline packs may facilitate restoration of blood flow and may delineate compromised segment of bowel. The bowel should also be inspected for pathologic lead points that should be removed. Finally, an appendectomy should be performed. If bowel resection is necessary, the patient can usually be managed with primary anastomosis.

Postoperatively, patients should be kept NPO until resolution of ileus. Use of postoperative antibiotics can be considered and should be used in cases of perforation or gross spillage.

Recurrence

The risk of recurrence is 5–7 % after enema reduction, almost two-third of which occur within the first 3 days of reduction. Recurrences have been noted, however, to occur as long as 3 years after the initial reduction. Recurrence after operative reduction is lower, usually around 2 %. Recurrent cases should initially be treated with attempts at nonoperative reduction. Due to the concern for occult malignancy, some clinicians recommend that surgical management be pursued in the following cases: (1) children with more than one recurrence with no previous surgery documenting absence of a lead point, (2) children older than 2 years whose initial reduction was nonoperative, and (3) children in whom a pathologic lead point is suspected (e.g., patients with polyposis syndromes). It should be noted that although initial recurrences are often successfully reduced

nonoperatively, the success rate diminished with repeated recurrences.

Finally, age is an important factor when considering the diagnosis and treatment of intussusception. Neonates, like adults, are more likely to have a surgical lead point for an intussusception—a diagnosis that is rare in this age group. Given that this likelihood is 60–75 %, that enema reduction has a low rate of success, and that the risk of perforation is higher, surgery is recommended once the diagnosis is made.

75
Hirschsprung's Disease

Jarod P. McAteer, Morgan K. Richards, and Adam B. Goldin

Introduction

Hirschsprung's disease (HD) is defined as **the absence of ganglion cells** over a variable length of intestine. *It always involves the rectum and extends proximally for an individually variable distance*. The disease is limited to the rectosigmoid colon in 75–80 % of cases, although it may extend to the transverse colon in up to 15 % of patients, to the entire colon and terminal ileum in 2–13 % of cases (total colonic aganglionosis), and in the rarest of cases may involve the entire intestinal tract to the duodenum. The lack of ganglion cells in the intestinal myenteric plexus and submucosal plexus leads to a *lack of normal relaxation* in the aganglionic segment, creating a functional obstruction and leading to significant dilation of the normal bowel proximal to this segment. The overall incidence is 1 in 5,000 live births, and 70–80 % of cases are in boys. The disease can be sporadic or familial, with 10–15 % of siblings of patients with total aganglionosis being afflicted with the same condition.

Although the pathologic characteristics of the disease are well described, the etiology is poorly understood. There are several theories as to the cause of the observed lack of ganglion cells. The first theory proposes that the disease is due to a failure of the proper caudal migration of neural crest cells during fetal development. Neuroblasts appear in the developing esophagus around week 5 of gestation and migrate down the intestinal tract outside of the circular muscle layer, reaching the anus around week 12. The longitudinal muscle forms over these cells, which differentiate into the myenteric plexus. The neuronal components of this plexus then migrate across the circular muscle layer into the submucosa and mucosa, further differentiating into the submucosal plexus. As these two plexi ultimately control bowel motility, failure of this migration at any point along this process will result in a variable length of affected bowel, beginning at the most distal point and extending over a caudal to cranial length.

A second theory proposes that the absence of ganglion cells is not due to a failure of migration, but rather to a failure of differentiation of neuroblasts secondary to changes in the local microenvironment. It has been shown that certain extracellular matrix proteins affect the path of neuroblast migration, while others affect maturation and differentiation. Intestinal ganglion cell immaturity, commonly termed *dysganglionosis*, can coexist with HD or be a component of other forms of neonatal intestinal obstruction.

Other theories point to the importance of genetics in the development of HD, given its familial clustering and its association with Down's and other syndromes. Several studies have suggested that the *RET* gene on chromosome 10 is responsible for the development of HD. Further work, however, has suggested that while genetics likely play a key role in disease etiology, the process is polygenic and complex. Other genes implicated in the pathogenesis of HD include the endothelin receptor B gene on chromosome 13 and the endothelin 3 receptor gene on chromosome 20. The complexity of the genetic contribution to this disease is reflected by findings suggesting that long-segment aganglionosis may have an autosomal dominant mode of inheritance, whereas short-segment disease shows a pattern more consistent with autosomal recessive inheritance.

Clinical Presentation

The most common presentation in neonates is failure to pass meconium within the first 24 h of life, a finding present in 90 % with the diagnosis of HD (Fig. 75.1). Other common symptoms include emesis and abdominal distension. While a large number of children present in the neonatal period with complete obstruction, some may suffer very few symptoms early on and present later in infancy or early childhood with chronic, progressive constipation. In such chronic cases, symptoms are often precipitated by changes in feeding, such as changes in formula type, or weaning from breast milk. It is not uncommon for these patients to present with progressive failure to thrive, anemia, and other signs of long-standing malnutrition. Often, if a rectal exam or rectal irrigation is

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_75, © Springer New York 2015

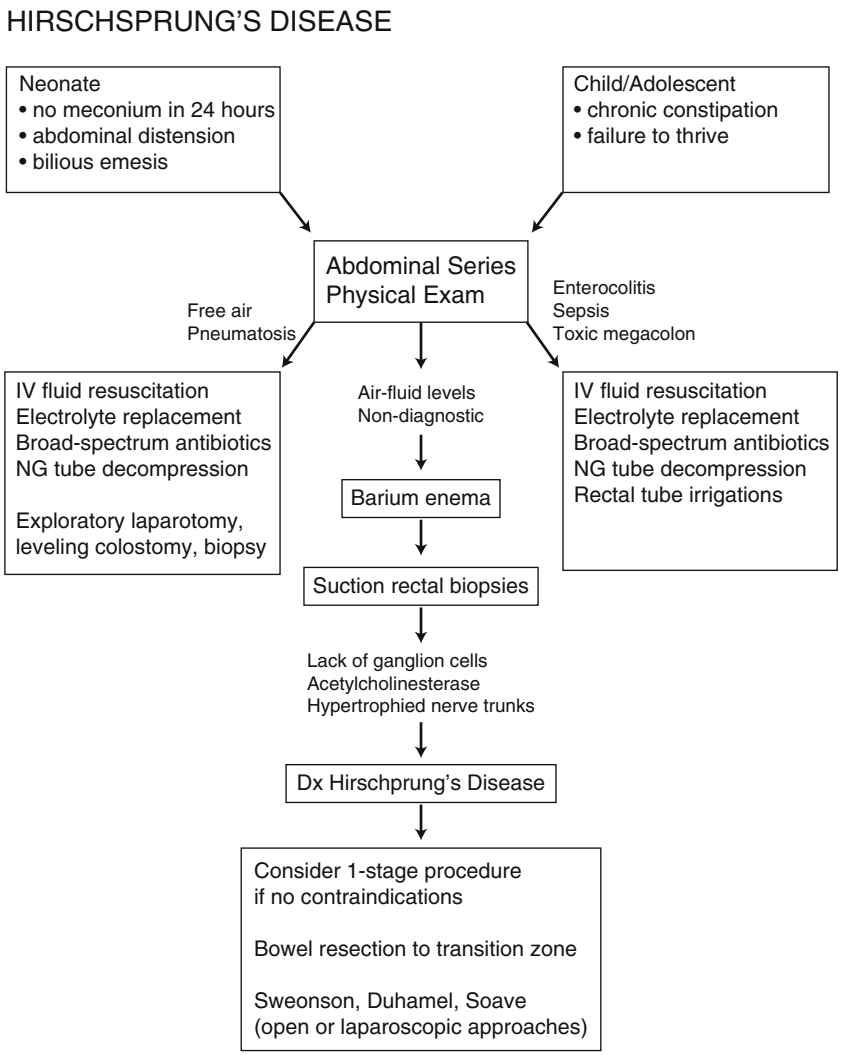

FIG. 75.1 Treatment algorithm for Hirschsprung's disease. *IV* intravenous, *NG* nasogastric, *Dx* diagnosis

performed, the meconium will pass with relief of symptoms for a short time, ultimately with return of distension thereafter. A small percentage of children may present with enterocolitis (15–50 %), which is manifested by diarrhea, mucosal sloughing, and sepsis. The etiology is not completely understood, although it is thought to be related to colonic distension secondary to fecal burden, leading to ischemia and bacterial translocation. In its most severe form, HD-associated enterocolitis may progress to toxic megacolon and septic shock. If not promptly treated, these patients may progress to pneumatosis and perforation, necessitating emergent operative intervention.

It is also important to be aware of other syndromes that are associated with the diagnosis of HD, as the presence of one should prompt a search for the others. These include trisomy 21 (seen in 3 % of HD patients), Waardenburg syndrome, cartilage-hair hypoplasia, and various cardiac anomalies. It is also important to consider and rule out HD in any neonate presenting with appendicitis, as aganglionosis is frequently the underlying cause of appendicitis in this age group.

Diagnostic Evaluation

Abdominal plain films are an important first step and can help to identify air-fluid levels consistent with bowel obstruction, air in a nondistended rectum, and, in severe cases, pneumatosis and pneumoperitoneum. In the absence of perforation, enterocolitis, or sepsis, patients should progress to barium enema as the next diagnostic step. Rectal irrigation and manipulation prior to contrast enema should be avoided, as they can result in a false-negative study result. Films that suggest the presence of HD typically show a narrow rectum with a cone-shaped portion of bowel extending up into a dilated segment of proximal bowel. Enema studies are

inconclusive in 10 % of cases and are often difficult to interpret in "short-segment" HD, as well as in total colonic aganglionosis. In cases of nondiagnostic contrast enema, it may be useful to reimage the patient in 24 h, as continued retention of contrast in the colon past that point is further suggestive of the diagnosis.

Some institutions utilize anorectal manometry as a screening tool for the diagnosis of HD. The diagnostic accuracy of this technique has been quoted as high as 85 %. A positive study is defined as absence of anorectal inhibitory response after balloon dilation of the rectum. The normal rectosphincteric reflex, which consists of a decrease in the resting pressure of the anal canal to such a distending stimulus, is typically thought to be mature by the 12th day of life. It has been noted, however, that the reflex may not be present in infants with gestational age less than 39 weeks or birth weight less than 2.7 g. As such, the utility of anorectal manometry in neonates is limited.

The gold standard for diagnosis of HD is rectal biopsy, and this should be pursued in all stable patients if the diagnosis is suspected based on the previously mentioned studies. Rectal biopsy is usually completed at the bedside via a suction rectal biopsy gun. Biopsies should extend to the depth of the submucosa. An absence of ganglion cells, the presence of hypertrophied nerve fibers, and an excess of staining for acetylcholinesterase are indicative of HD.

Operative Therapy

The initial management of a child with HD and enterocolitis includes IV fluid resuscitation and electrolyte correction, broad-spectrum antibiotics, and NG decompression. Patients may benefit from rectal irrigation (10–20 mL/kg several times per day) and decompression via a rectal tube. In cases of enterocolitis, all of these measures are essential in stabilizing the patient prior to going forward with definitive HD workup. However, in cases of refractory sepsis due to toxic megacolon or perforation, operative intervention should not be delayed beyond an initial period of resuscitation.

The ultimate goals of operative therapy for HD are to remove the aganglionic bowel and to restore continence and normal function. Historically, this was achieved in two stages, even in elective cases. Although some still advocate a two-stage procedure (beginning with colostomy and then a pull-through at a later time) in infants diagnosed in the first few months of life, surgeons are increasingly employing single-stage procedures even in the neonatal period. Absolute indications for a diverting colostomy (or even loop colostomy in the most unstable infants) include severe malnutrition, massive proximal bowel dilation, unresolved enterocolitis or toxic megacolon, perforation, total colonic aganglionosis, or lack of pathology support. Surgeon comfort level should also factor into the decision, and it should be remembered that a two-stage procedure is always a safe and reasonable option if doubt exists.

The next important consideration in operative management of HD is the amount of bowel to be resected. The goal is to remove all diseased bowel including the transition zone between dilated and decompressed bowel. The transition zone cannot be reliably defined by radiographic criteria, as 10 % of HD patients have no radiographic transition zone, and another 8 % have a pathologic transition zone proximal to the radiographic transition zone. For this reason, *intraoperative frozen sections are an important adjunct to management*. It is also important to note that the transition zone itself may be deceiving. A single biopsy on one side of the bowel may not accurately identify the transition zone, as it is frequently not symmetrical around the circumference of the bowel. Further, even with the appearance of normal-appearing ganglion cells, functional dysganglionosis may persist for another 10–15 cm proximal to the point at which ganglion cells first appear. Pull-through using this segment may result in residual obstructive symptoms. As such, many experts now advocate resecting proximal to the first normal biopsy, with recommendations ranging from 2 to 15 cm.

Once the aganglionic segment has been resected, definitive treatment for HD has generally been achieved by one of three procedures: the **Swenson**, the **Duhamel**, and the **Soave** pull-throughs. The *Swenson* procedure was first described in 1948 and involves complete resection of the aganglionic bowel to within 1.5 cm of the dentate line anteriorly and 1.0 cm posteriorly, thus preserving the anal sphincter. A sutured end-to-end anastomosis is created externally between the normal proximal end of bowel and the everted rectal stump. The *Duhamel* procedure was first described in 1956 and modified over 10 years. In this procedure, the aganglionic rectum is not resected, and the proximal functional bowel is brought into the pelvis posteriorly. The two lumens are then unified using a long GIA stapler, creating a single common rectal reservoir. Finally, the *Soave* procedure, described in 1964, involves resection of a tube of mucosa and submucosa from the aganglionic bowel to within 1.0 cm of the dentate line. The normal proximal bowel is then brought through the remaining muscular sleeve, and an anastomosis is created at the anus. The proposed advantage of the Duhamel and Soave procedures is their avoidance of dissection along the anterior rectal wall, which may disrupt pelvic nerves responsible for continence and sexual function. Laparoscopic techniques have become increasingly popular, and each of the three pull-through procedures have been successfully performed using laparoscopy.

Complications

In addition to complications such as wound infection, patients with HD may experience other complications after surgery. One of the most common is persistent constipation or

obstructive symptoms. If severe, these symptoms should prompt diagnostic evaluation and repeat rectal biopsies, as they may be indicative of anastomotic stricture or residual aganglionosis (or transition zone bowel). In these cases, patients should be managed with a repeat pull-through procedure.

Even after definitive surgery, children are still at an increased risk for the development of enterocolitis. The reason for this is unknown, but it is important to remember this possibility in any child who has the diagnosis of HD and presents with bowel complaints. As before, these children should be treated with IV fluid resuscitation and electrolyte correction, NG decompression, bowel rest, and broad-spectrum antibiotics. These children may also require rectal irrigation or decompression. If a child does not improve with these maneuvers, one must consider returning to the basic principles of total parenteral nutrition and diverting colostomy.

76
Necrotizing Enterocolitis

Loretto Ann Glynn

Introduction

Necrotizing enterocolitis (NEC) is the most common surgical emergency encountered in the neonatal intensive care unit, occurring in 1–3/1,000 live births. Over 90 % of affected infants are premature and weigh less than 2,000 g. As advances in neonatal care have led to increased survival rates of infants of even smaller birth weight and earlier gestational age, the incidence of NEC has increased. The mortality for infants with NEC is 10–50 % with a decrease of surgical mortality to 20–50 %.

Pathophysiology

The pathophysiology of NEC has been and continues to be studied extensively. Research findings have supported the concept that NEC is the result of intestinal ischemia, colonization by pathogenic bacteria, and protein substrate in the intestinal lumen (Fig. 76.1). Initially, the infant suffers a subclinical insult due to hypoxia, poor perfusion, or infection. Bacteria bind to the injured mucosa, inciting an escalating inflammatory response leading to mucosal injury. Many inflammatory mediators have been identified, including interleukin-18, interleukin-12, TNF, and platelet-activating factor. A lack of protective agents such as epidermal growth factor and nitric oxide synthase contributes to the development of NEC. The immature intestine has a maladaptive vasoconstrictive response to hypoxia, ineffective peristalsis, compromised immunologic function, and a decrease in pepsin, gastric acid, and mucins. These factors augment the pathologic process. NEC is most commonly diagnosed in the second week of life, after colonization of the intestinal tract has occurred. Studies have suggested a relationship between the transfusion of packed red blood cells and the development of NEC in otherwise stable neonates who were feeding and growing. This may be due to an immunologic response in the intestinal mucosa to the blood group antigens. Further research in this area is necessary to understand this relationship.

Clinical Presentation

NEC typically presents in the second week of life, most commonly affecting premature, low-birth-weight infants. Additional risk factors include the use of antenatal or postnatal indomethacin, cardiac shunts, umbilical artery catheters, and cocaine exposure. Many cases of NEC have occurred in infants who have had early enteral feeding. The early signs of NEC can be nonspecific, including apnea, bradycardia, and temperature instability. More specific findings include blood in the stool (up to 63 % of cases), feeding intolerance, and emesis. *The most common finding is abdominal distention frequently accompanied by abdominal tenderness and rigidity*. Advanced cases can present with abdominal wall erythema, ecchymosis, and edema.

Laboratory Findings

Infants with NEC can have an elevated white blood cell count or absolute neutropenia. Neutropenia is associated with fulminate NEC. Thrombocytopenia and metabolic acidosis are common and are associated with a poor prognosis. Research has focused on identifying early markers of disease that may identify infants at risk for developing NEC, in the hopes that properly identifying them would result in a decrease in unnecessary cessation of feeding, laboratory tests, and radiographs. Thus far, none of the markers studied have proven to have the specificity and sensitivity to be a useful diagnostic tool.

Radiographic Findings

Radiographic studies are important in the diagnosis and staging of NEC. According to Bell, stage 1, or suspected NEC, is characterized by an ileus on plain abdominal radiographs, along with clinical findings of abdominal distention, feeding intolerance, and vomiting. Stage 2 includes

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_76, © Springer New York 2015

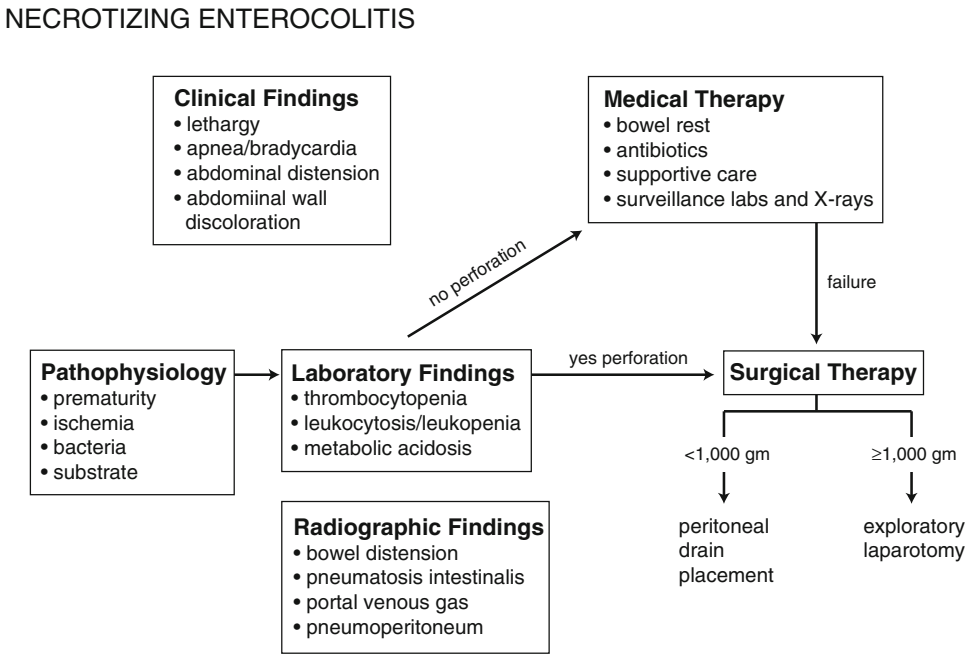

Fig. 76.1 Treatment algorithm for necrotizing enterocolitis

gastrointestinal bleeding and radiographic findings of pneumatosis intestinalis (70–80 % of cases) and portal venous gas (25 % of cases). Stage 3, advanced NEC, is characterized by pneumoperitoneum on abdominal radiographs and sepsis clinically.

Ultrasound has been shown to be as effective as abdominal radiographs at identifying pneumatosis intestinalis, portal venous gas, and pneumoperitoneum. In addition, ultrasound can determine the presence of intra-abdominal fluid, bowel wall thickness, and bowel perfusion.

Medical Management

Management of NEC is based on Bell's stage. Stage 1, suspected NEC, is treated with a brief period of bowel rest, decompression with a nasogastric tube, broad-spectrum antibiotics, and supportive care. Antibiotic regimens should cover gram-positive bacteria, aerobic gram-negative bacteria, and anaerobes. The most common regimen is ampicillin, gentamicin, and clindamycin. Infants with suspected NEC are monitored with serial abdominal exams, abdominal radiographs, complete blood counts, and blood gases. If the infant does not develop stage 2 NEC after 72 h, then treatment can be discontinued. If the infant develops pneumatosis intestinalis (Bell's stage 2), then treatment should continue for 7–10 days. If pneumoperitoneum develops (Bells' stage 3), then surgical intervention is required.

Surgical Management

The two main indications for surgical management of NEC are bowel perforation and failure of medical management. Perforation is diagnosed by pneumoperitoneum on abdominal radiograph or paracentesis positive for stool or bile. Failure of medical management can be defined by oliguria, hypotension, metabolic acidosis, thrombocytopenia, respiratory failure, portal venous gas, persistently dilated bowel loops, and abdominal wall erythema. Up to 50 % of infants with NEC will require operative management.

Findings at laparotomy can show involvement of any portion of the intestine, most commonly both small and large intestines in segments. Fifteen to twenty percent will have NEC totalis, defined as involvement of >75 % of the intestine. The goal of exploratory laparotomy includes the limited resection of necrotic bowel and creation of stomas. Early ostomy reversal at 6 weeks post-laparotomy is well tolerated. Primary anastomosis is seldom possible due to inflammation and bleeding.

Pan-involvement is associated with a poor outcome. Survival often results in short bowel syndrome requiring long-term parenteral nutrition and is associated with a high mortality rate. Therefore, resection of bowel should be limited to frankly necrotic bowel. Several surgical approaches have been utilized to aid in the preservation of bowel length, including the use of second-look laparotomy to allow questionable bowel to be evaluated for viability, preserving as

much bowel length as possible, and also the "clip-and-drop back" technique, where necrotic bowel is removed and ends are clipped and placed back into the abdomen with restoration of bowel continuity at 48–72 h later.

Primary peritoneal drainage can be an option to manage bowel perforation in infants weighing less than 1,000 g. Penrose drains are placed into the abdomen through bilateral lower quadrant incisions. Drains are left in place for resuscitation and medical therapy. If the infant's condition does not improve, a salvage laparotomy can be performed. Survival for infants requiring laparotomy after failure to improve with peritoneal drains is associated with a high mortality rate. Infants who improve after drain placement can be managed without laparotomy in the acute setting. However, they frequently go on to have bowel obstruction that requires laparotomy later in their course. Primary peritoneal drainage has not been proven to be superior to laparotomy in prospective trials. However, there seems to be a survival advantage for extremely low-birth-weight infants when compared to laparotomy in the acute setting.

Strictures may develop in 20–30 % of infants successfully treated medically and up to 50 % of those treated with laparotomy. Strictures occur most frequently in the colon and typically present at 4–6 weeks after acute NEC. Signs include abdominal distention, feeding intolerance, blood in stool, and vomiting. The diagnosis can be confirmed with a barium enema. Treatment requires exploratory laparotomy and resection of the stricture.

Outcome

Unfortunately, the incidence of NEC is increasing. Even with the advances in surgical and neonatal intensive care, the survival after NEC is not increasing. Mortality is 20–50 % with very-low-birth-weight infants and those with the lowest gestational age having the highest mortality. Short bowel syndrome occurs in 10 % of survivors, resulting in considerable morbidity and cost of care.

Long-term follow-up has shown that while the majority of children ages 5–10 years were enrolled in school, 14 % had developmental delay. There may be confounding factors in these patients, such as the presence of intraventricular hemorrhage. Additional studies are needed to learn more about the true relationship between NEC and poor neurodevelopmental outcome.

Part VIII
Urology

77
Hematuria

Michael P. Hoeh and Kalyan C. Latchamsetty

Introduction

Hematuria may originate from any site along the urinary tract and, whether gross or microscopic, may be a sign of serious underlying disease, including malignancy. A urologic cause for hematuria is often not identified (61 % in a series of more than 1,900 patients referred to a hematuria clinic). However, given the risk of a potentially serious underlying problem, a genitourinary evaluation is recommended. Hematuria may be grossly visible (macroscopic hematuria) or only detectable on microscopic urine examination (microscopic hematuria). Microscopic hematuria is defined as three red blood cells (RBCs) per high power field (HPF) or greater on microscopic evaluation of the urinary sediment from two of three properly collected urinalysis specimens.

Indications for Urologic Workup

Cancer is more common in patients with gross hematuria (23 %) than in patients with microscopic hematuria (5 %). The literature agrees that gross hematuria warrants a thorough diagnostic evaluation (Fig. 77.1); in contrast, microscopic hematuria may be an incidental finding, and whether physicians should test for hematuria in asymptomatic patients remains controversial. The American Urological Association (AUA) currently recommends that nontraumatic adult patients with at least *two of three* properly collected urine specimens with three or more RBCs per HPF should undergo a complete hematuria workup (Table 77.1). The AUA also recommends that if a patient has one or more risk factors (Table 77.2) for significant urologic disease, he/she should be referred for a full urologic evaluation after *one* properly performed urinalysis documenting the presence of at least three red blood cells per high power field.

History and Physical Exam

An accurate history and physical examination are essential components of the diagnostic workup; collectively, they are extremely helpful in elucidating the cause of the hematuria. A proper history should focus on the duration and character of the hematuria, associated symptoms, age of patient, past medical history, history of urologic procedures, history of any of the risk factors listed in Table 77.2, and history of trauma or recent vigorous activity. Irritative voiding symptoms (i.e., urgency, frequency, and dysuria) are suggestive of an infection. Hematuria in sexually active females is most often caused by cystitis secondary to a urinary tract infection. Hematuria associated with severe flank pain is suggestive of nephrolithiasis, whereas painless hematuria is concerning for a tumor or renal parenchymal disease. Lower urinary tract symptoms, such as weakened stream and hesitancy, can indicate an obstructive process that may be secondary to benign prostatic hypertrophy (BPH). The prostatic urethra in a patient with BPH is friable and has a tendency to bleed; moreover, patients with an obstructive process are more likely to develop cystitis. Cyclic hematuria in women that is most prominent during and shortly after menstruation may suggest endometriosis of the urinary tract.

When evaluating gross hematuria, the timing of the hematuria can be very helpful in isolating a source of bleeding. Gross hematuria may be classified as initial, total, or terminal, depending on when the hematuria appears during micturition. Initial hematuria is indicative of urethral bleeding distal to the external urinary sphincter. Total hematuria suggests bladder or kidney bleeding. Finally, terminal hematuria is indicative of urethral bleeding proximal to the external urinary sphincter (i.e., prostate or bladder neck).

A complete physical examination should be performed with special attention focused on the patient's vital signs, abdominal exam, and genitourinary exam. Pyelonephritis may

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_77, © Springer New York 2015

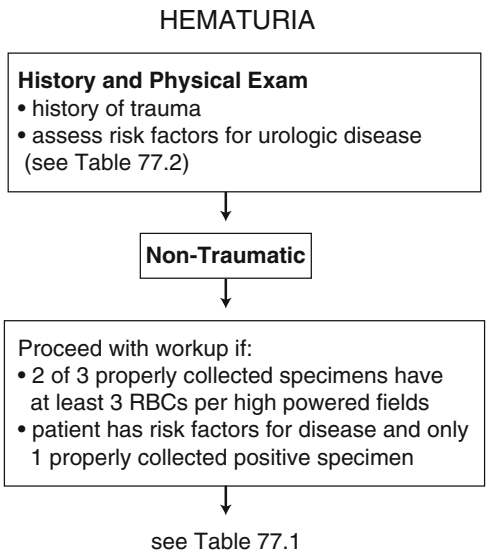

FIG. 77.1 Treatment algorithm for hematuria. RBCs, red blood cells

TABLE 77.1. Non-traumatic hematuria workup for adults.

History and physical exam
Urinalysis (confirm a positive dipstick with a microscopic examination)
Continue hematuria workup if any of the following criteria are present
Gross hematuria or > 100 RBCs per HPF
Microscopic hematuria or at least three RBCs per HPF in two of three properly collected urine specimens
Microscopic hematuria in at least one properly collected urine specimen in patients with risk factors for GU disease (as defined in Table 77.2)
Urine culture
If UA or culture indicates infection, recommend to treat for UTI and recheck in ~6 weeks to determine if hematuria persists. If so, then continue workup as listed below
Urine cytology
Blood tests – BUN and creatinine, check PSA in older men. Consider CBC with differential, platelets, PT, PTT, sickle cell test in African Americans
Upper tract imaging (choose from either option listed below)
CT urogram – diagnostic test of choice
Renal ultrasound with either IVP or retrograde pyelogram
Cystoscopy
If blood arises from ureter, consider ureteroscopy

TABLE 77.2. Risk factors for significant urologic disease in patients with microscopic hematuria.

Smoking history
Occupational exposure to chemicals or dyes (benzenes or aromatic amines)
History of gross hematuria
Age > 35 years
History of urologic disorder or disease
History of irritative voiding symptoms
History of recurrent urinary tract infections
Analgesic abuse
History of abdominal-pelvic radiation

present with costovertebral angle (CVA) tenderness as well as an elevated temperature (>101 °F). Nephrolithiasis associated with ureteral obstruction often presents with acute CVA tenderness, which may radiate to the lower abdominal/genital region with or without nausea and vomiting. An enlarged prostate on digital rectal exam may indicate BPH, whereas an exquisitely tender prostate on exam suggests prostatitis.

Urinalysis and Urine Dipstick

Urine dipstick and microscopic examination should be performed on all patients with hematuria. The urine dipstick assesses the presence of hemoglobin that is either within the urinary RBCs or free in the urine. A positive test is indicated by a specific color change on the dipstick; this oxidation reaction is catalyzed by hemoglobin. It is important to note that false positives may be the result of myoglobin as well as oxidizing contaminants such as Betadine. Urinary dipsticks have a sensitivity of 91 % in detecting asymptomatic microscopic hematuria; their specificity is limited, however, and has been reported to be as low as 65 %. As a result, each positive dipstick needs to be confirmed with a microscopic evaluation.

Renal parenchymal disease can sometimes present as hematuria. When significant proteinuria, RBC casts, renal insufficiency, and/or predominance of dysmorphic RBCs are found in the urinalysis, a nephrologist should be consulted.

Cytology

The sensitivity of urine cytology is greatest for bladder cancer (approximately 90 %). By comparison, sensitivity for upper tract carcinoma is limited, with the reported false-negative rate overall being 65 %. Nevertheless, this is a simple test that may provide valuable information in the hematuria workup.

Imaging

The initial evaluation of hematuria should include urologic imaging. Imaging can be used to detect renal cell carcinoma, urothelial cell carcinoma (UCC), urolithiasis, and renal infection. The diagnostic yield of imaging studies in adults increases with age and is typically higher for gross hematuria than for microscopic hematuria. A number of modalities are available to evaluate the genitourinary tract among patients with unexplained hematuria. These include conventional radiography, intravenous pyelography (IVP), retrograde pyelography, ultrasonography, magnetic resonance imaging, magnetic resonance urography, conventional CT scanning, and multiphasic CT urography.

Most clinicians consider multiphasic CT to be the preferred initial imaging modality in most patients with unexplained hematuria. This modality may be particularly

indicated in those with an increased risk of genitourinary malignancy or disease. The combination of multiphasic CT and cystoscopy, which together provide a complete evaluation of the genitourinary system, should be performed in almost all patients with unexplained hematuria. Patients with diminished renal function can undergo an MRI or a combination of renal ultrasound and retrograde pyelogram.

Cystoscopy

A careful inspection of the urethra and bladder with a cystoscope is a vital aspect of the workup for hematuria. In those instances where an individual presents with asymptomatic microscopic hematuria and does not have any of the risk factors for bladder cancer (see Table 77.2), a cystoscopy may not be necessary if the aforementioned workup is otherwise negative. Cystoscopy may be used to establish the diagnosis of bladder cancer, assess whether or not muscle invasion is present, and provide initial therapy for non-muscle-invasive lesions.

In the instances in which hematuria is documented but no identifiable cause can be found through the aforementioned workup, close follow-up is indicated.

78
Testicular Mass

Christopher L. Coogan, Jonas S. Benson, and Ryan L. Steinberg

Differential Diagnosis

The differential diagnosis of a *painless* testicular mass includes a spermatocele, hydrocele, varicocele, inguinal hernia, and testicular tumor (Fig. 78.1). Testicular torsion, epididymitis, trauma, and incarcerated inguinal hernia can also cause a testicular mass but typically are associated with pain. A tumor that hemorrhages or grows rapidly may cause acute testicular pain, although this occurs infrequently. Testicular tumors are relatively rare and constitute only 1–2 % of all male malignancies. The median age of diagnosis is 33 years. They are the most common tumors in men aged 20–40 years and are currently one of the most curable solid neoplasms.

History and Physical Examination

A thorough history and physical examination should be performed on all patients. The usual presentation of a testicular tumor is a painless mass; approximately 30–40 % of patients will complain of a dull ache or heaviness in the scrotum. Symptoms related to metastatic disease are the presenting complaint in 10–20 % of patients and include flank pain, palpable mass, and lower extremity swelling. Gynecomastia is seen in 2 % of patients as a result of either hormone secretion from the tumor or androgen deficiency. Risk factors for testicular tumors are cryptorchidism (4–6 times more likely), family history of testicular cancer, personal history of testicular cancer, and intratubular germ cell neoplasia.

Physical examination of the testes is performed by first examining the normal testicle. A hard or fixed area within the testicle is considered cancer until proven otherwise. Atrophy of the affected or contralateral testicles is common. One can usually differentiate between a testicular tumor, varicocele, hydrocele, spermatocele, epididymitis, and testicular torsion by history and physical examination alone. *Varicocele* is the dilation of the pampiniform venous plexus; it occurs more often on the left side and will increase in size when one moves from supine to standing position or with Valsalva. *Hydrocele* is a fluid collection within the tunica vaginalis, although normally benign, 5–10 % of testicular cancers have an associated reactive ipsilateral hydrocele. *Spermatocele* is the cystic dilation of an epididymal tubule. Both hydrocele and spermatocele can be detected by transillumination of the scrotum. A scrotal ultrasound may be needed to establish an accurate diagnosis, especially in patients with large hydroceles, which can make testicular palpation difficult.

Laboratory Tests

Urinalysis may be helpful and may differentiate epididymitis from a tumor. The most definitive studies are the tumor markers alpha fetoprotein (AFP) and the beta subunit of human chorionic gonadotropin (B-HCG). Tumor markers should be drawn promptly for diagnostic, prognostic, and therapeutic purposes. AFP is elevated in patients with embryonal cell carcinoma, teratocarcinoma, yolk sac tumors, or mixed tumors. AFP has a serum half-life of 5–7 days. B-HCG is elevated in all patients with choriocarcinoma, 50 % of embryonal cell carcinomas, and 5–10 % of seminomas. B-HCG has a half-life of 24–36 h. Lactate dehydrogenase (LDH) is another commonly measured tumor marker that is not specific to testicular cancer but can be an indicator of tumor burden. LDH has a half-life of 24 h. Ten to fifteen percent of nonseminomatous germ cell tumors will not exhibit tumor marker elevation. In addition, *the majority of seminomas do not exhibit any tumor marker elevation*. The failure of tumor markers to return to normal levels post-orchiectomy is indicative of metastatic disease, and these patients should receive chemotherapy.

Ultrasound Evaluation

A scrotal ultrasound is the gold standard imaging modality to evaluate testicular masses of uncertain etiology. Testicular tumors are visualized sonographically as hypoechoic lesions

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_78, © Springer New York 2015

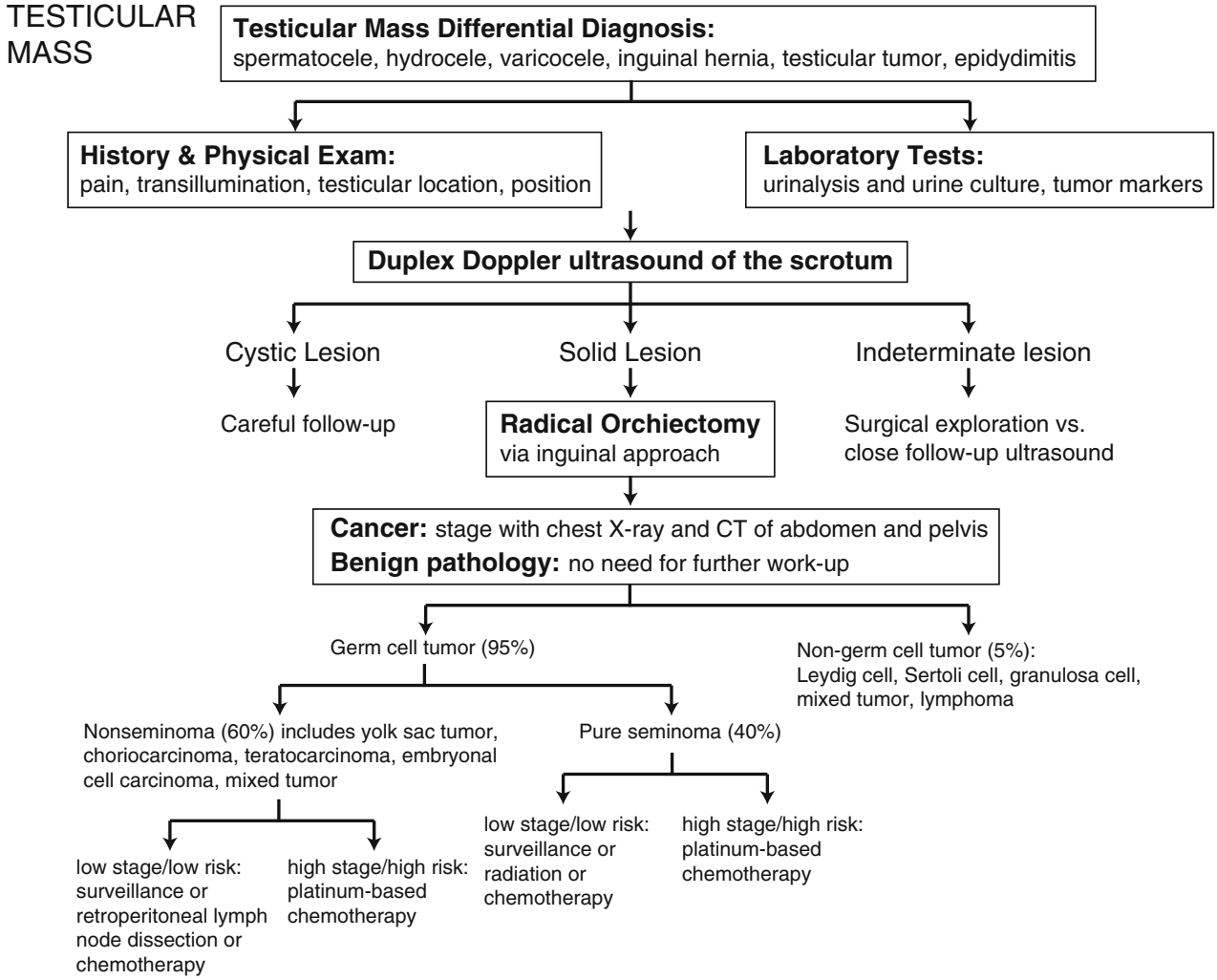

FIG. 78.1 Testicular mass treatment algorithm

arising within the tunica albuginea. Epididymitis, torsion, hydrocele, spermatocele, and varicocele may also be diagnosed by ultrasound.

Orchiectomy

Removal of a testicular tumor is performed through an **inguinal approach** to permit early clamping and high removal of the spermatic cord. Orchiectomy should **not** be performed through a scrotal approach as the scrotal skin has a different lymphatic drainage pattern than the testicle. The orchiectomy specimen provides a definitive histological diagnosis of a testicular tumor and has therapeutic benefit to the patient.

Cancer

If cancer is confirmed, staging studies for patients should include a chest X-ray and computed tomography (CT) scan of the abdomen and pelvis. Testicular cancer generally spreads first to the retroperitoneal lymph nodes with the exception of choriocarcinoma; this area must be examined with the CT scan. A CT scan of the chest is performed in select patients. Ninety-five percent of testicular tumors are of germ cell origin; they are further divided into two general classes: seminoma and nonseminomatous germ cell tumors. Long-term survival of patients with metastatic germ cell tumors is 80–90 %. Testicular lymphoma is the most common testicular tumor in men greater than 50 years of age.

Seminoma is the most common testicular tumor in adults and accounts for 40–60 % of testicular tumors. Twenty to 25 % of seminomas present with metastatic disease. Low-stage pure seminoma (no retroperitoneal adenopathy or low-volume retroperitoneal disease) is treated with orchiectomy and surveillance or chemotherapy or radiation therapy to the retroperitoneal lymph nodes. High-stage seminoma is treated with platinum-based combination chemotherapy.

Nonseminomatous germ cell tumors include embryonal cell carcinomas, teratomas, choriocarcinomas, and yolk sac tumors alone or in combination. Fifty to 70 % of nonseminomatous germ cell tumors present with metastatic disease. Testicular tumors that contain both seminoma and nonseminomatous germ cell tumors are treated as nonseminomatous germ cell tumors. The incidence of positive nodes in patients without evidence of adenopathy on CT scan is 30 %. Low-stage/low-risk disease may be treated with orchiectomy followed by surveillance or retroperitoneal lymph node dissection or chemotherapy. Patients with high-stage/high-risk disease are treated with platinum-based combination chemotherapy.

Non-germ cell tumors constitute only 5 % of testicular neoplasms. This group includes Leydig cell tumors, Sertoli cell tumors, granulosa cell tumors, gonadoblastoma, mixed tumors, and lymphoma. Radiation and chemotherapy are ineffective against metastatic Leydig and Sertoli tumors. Gynecomastia and other feminizing characteristics are common presenting symptoms with these tumors due to peripheral androgen conversion.

79
Painful Scrotum

Christopher L. Coogan, Jonas S. Benson, and Ryan L. Steinberg

Differential Diagnosis

The differential diagnosis of a painful scrotum includes epididymitis, orchitis, testicular torsion, testicular appendage torsion, Fournier's gangrene, incarcerated inguinal hernia, hydrocele, varicocle, and trauma. Hydrocele and varicocele do not typically present with pain.

History and Physical Examination

The etiology of scrotal pain can usually be determined on the basis of history and physical examination (Fig. 79.1), although laboratory tests, radiological examination, and occasionally scrotal exploration may also be necessary to confirm the diagnosis. The duration of pain (acute vs. chronic) and age of the patient (child/adolescent vs. adult) are particularly helpful in narrowing the differential. Spermatic cord torsion may occur at any age but primarily occurs in neonates and adolescents. This condition usually presents with the sudden onset of severe pain. A history of anatomic abnormalities such as a bell-clapper deformity (partial or complete failure of fusion of the tunica vaginalis along the epididymis resulting in incomplete attachment of the testis/epididymis to the scrotum), family history of torsion, and cryptorchidism are risk factors. Conversely, epididymitis and orchitis typically occur in adolescents and adults and have a more gradual onset of pain. A history of urinary tract infection, vomiting, dysuria, trauma, sexually transmitted disease, diabetes, previous swelling or pain, or recurrent infection are risk factors for epididymitis.

Examination of a painful scrotum can be extremely difficult. The unaffected side should always be examined first. The consistency, texture (nodularity), and position of the testicle within the scrotum and the ability to transilluminate the scrotum (hydrocele and spermatocele) are all helpful in determining the etiology. The absence of a cremasteric reflex (stroking of the medial thigh resulting in scrotal elevation) is often seen in torsion, as well as a high-riding testicle where the testicle lies horizontally in the proximal part of the scrotal sac. Further, manual elevation of a torsed testicle will not relieve pain (negative Prehn's sign) as is the case with epididymitis. Careful examination of scrotal skin is also important as it may reveal a discrete darkened area (blue dot sign) consistent with torsion of the testicular appendage, dusky/necrotic skin consistent with Fournier's gangrene, or signs of trauma. In epididymitis, the spermatic cord and tail of the epididymis are tender and swollen on palpation. Rarely, testicular tumors will present with testicular pain and should always remain in the differential diagnosis.

Laboratory Tests

Laboratory evaluation should include a urinalysis and urine culture in all patients and a complete blood count (CBC) in selected patients. Pyuria indicates a urinary tract infection and is also seen in some patients with epididymitis and orchitis. The urinalysis is usually normal in torsion. Profound leukocytosis in a patient with fever and severe testicular pain may necessitate hospital admission and intravenous (IV) antibiotics.

Radiography

The two modalities utilized in the evaluation of scrotal pain include a radioisotope scan and duplex ultrasound. The former is primarily of historic value as it is not as quickly available as ultrasonography. It is performed by IV infusion of technetium-99 into the patient and measuring isotope uptake by the testicles using a gamma camera. A torsed testicle has decreased blood flow and thus demonstrates a defect in isotope uptake. Duplex ultrasonography of the scrotum is the gold standard radiographic modality and can evaluate the absence of blood flow, presence of a testicular mass, increased epididymal blood flow (as seen in epididymitis), and the presence of intestinal contents in the scrotum (inguinal hernia).

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_79, © Springer New York 2015

PAINFUL SCROTUM

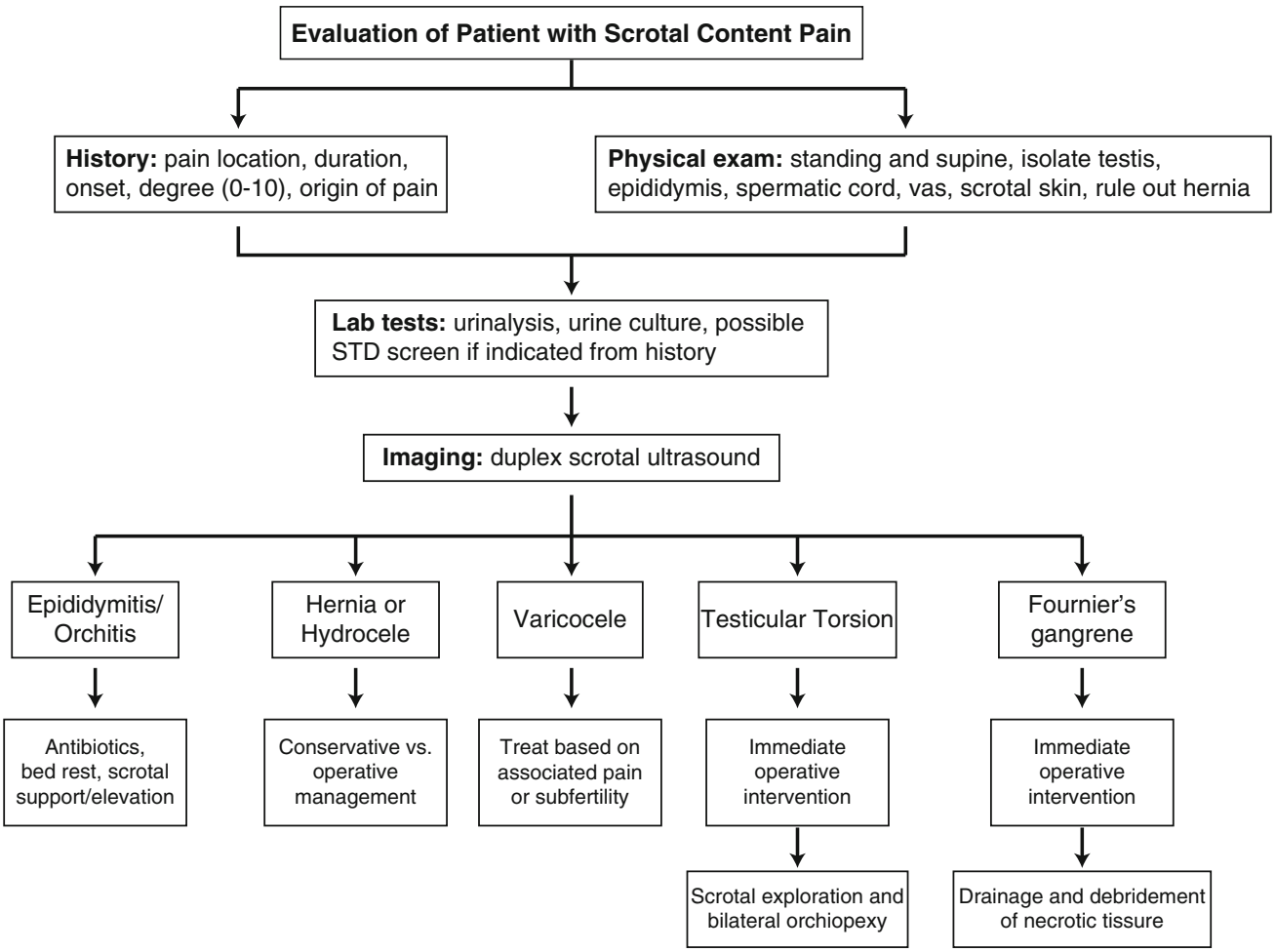

Fig. 79.1 Treatment of painful scrotum. *STD* sexually transmitted disease

Epididymitis/Orchitis

Epididymitis and orchitis can be seen in all age groups. It is typically related to a urinary tract infection or congenital anomaly in children, sexually transmitted infection in men between the ages of 18–35, and urinary stasis or catheterization in elderly men. It is due to the spread of infection from the prostate, bladder, or urethra to the testicle or epididymis. The onset of pain is usually gradual and, in cases of epididymitis, localized to the posterior aspect of the testicle. The testis and epididymis are firm, tender, erythematous, and swollen; patients may also have fever, dysuria, and leukocytosis. A duplex ultrasound or isotope scrotal scan will reveal increased blood flow to the affected testes. Patients are treated with IV or oral antibiotics, scrotal elevation, warm or cold compresses, and bed rest.

Testicular Torsion

Testicular torsion may occur at any age. In neonates the mechanism is usually extravaginal torsion, which is the twisting of the spermatic cord with all its contributing structures of the vas, vessels, processus vaginalis, and investing fascias. Adolescents and adults usually have an intravaginal torsion, which involves only the testis without its investing tunica vaginalis. The onset of pain is usually sudden, and patients may complain of previous episodes. The pain will often radiate to the groin and lower abdomen and may be associated with nausea and vomiting. Physical examination will reveal a very high-riding, diffusely swollen, and exquisitely tender testicle. The cremasteric reflex is often absent and patients will not have relief of the pain with testicle

elevation (Prehn's sign). A duplex ultrasound or radioisotope scrotal scan will reveal diminished or absent blood flow to/isotope uptake by the testicle. Patients with testicular torsion should undergo immediate scrotal exploration and orchiopexy of both testicles, as there is an increased risk of torsion occurring in the contralateral testes. The incidence of infarcted testicles increases significantly after 6 h of spermatic cord torsion.

Fournier's Gangrene

Patients with Fournier's gangrene (scrotal gangrene or necrotizing fasciitis of the perineum) often present with signs of external genital cellulitis (pain, erythema, warmth) as well as systemic illness (fever, chills, generalized malaise). Risk factors include diabetes, ethanol abuse, perineal trauma, recent instrumentation/surgery, paraphimosis, malnutrition, periurethral urine extravasation, and perirectal/perianal infections. The onset of symptoms may be insidious, especially in patients with a history of diabetes and ethanol abuse. Physical examination is diagnostic and usually reveals crepitus, necrosis, or purulent drainage on the penis and scrotum in addition to the features described previously. An abdominal x-ray, CT scan, or scrotal ultrasound may be beneficial to demonstrate free subcutaneous air. Treatment consists of hemodynamic stabilization, administration of broad-spectrum IV antibiotics, and immediate surgical debridement of all necrotic and devascularized tissue.

Incarcerated Inguinal Hernia

Incarcerated inguinal hernias may also cause sudden scrotal pain. Physical examination is often diagnostic, but occasionally CT scan and/or ultrasound is necessary for definitive diagnosis. Depending on the clinical scenario, reduction may be attempted. If successful, repair is generally undertaken during the same hospitalization. If reduction was unsuccessful or viability of herniated bowel is in question, emergent surgery is indicated.

Hydrocele

Hydroceles are defined as the presence of a persistent processus vaginalis. Physical examination usually demonstrates a swollen but nontender scrotum that will transilluminate, although a hydrocele that suddenly increases in size may result in testicular pain. If palpation of the testes is difficult, ultrasonography of the scrotum is necessary to ensure the absence of any other pathology. The majority of hydroceles do not require treatment other than reassurance and careful examination. Hydroceles may "communicate" with the peritoneal cavity through a hernia sac, or they may be "noncommunicating."

Varicocele

Varicocele represents a dilation of the spermatic cord veins also known as the pampiniform plexus, which has been referred to as a "bag of worms." The incidence of varicoceles is about 15 % in the general population of adult males and may result in subfertility. There is a marked predominance of left-sided varicoceles given the difference in venous drainage from the gonadal vessels. The left gonadal vein typically drains into the left renal vein, while the right gonadal vein drains directly into the IVC. Patients may report heaviness or a dragging scrotal sensation. Physical examination of a varicocele should be performed in both the supine and standing position to allow dilation of the pampiniform plexus. While supine, palpation of the spermatic cord at rest and during a Valsalva maneuver allows for grading of the varicocele. Varicoceles are rarely symptomatic and seldom require treatment except in the presence of subfertility or pain.

Part IX
Gynecology

80
Acute Gynecologic Pelvic Pain

Joseph M. Maurice and Bruce A. Rosenzweig

Introduction

Acute pelvic pain is a common complaint among women. A detailed description of the pain should be obtained with attention to its timing, duration, characterization, and modifying factors. A complete gynecological history should include the patient's last menstrual period, a description of the menstrual cycle, use of contraceptives, sexual history, sexually transmitted infections (STI), and previous self-therapies (Fig. 80.1). Workup should include a pregnancy test in all reproductive-age women. The categories of acute pelvic pain are divided by anatomic structure; however, some conditions may involve multiple pelvic organs.

Vulva

Acute pelvic pain of the vulva is seen after **blunt trauma**, namely, a straddle injury, obstetrical laceration, sexual assault, motor vehicle accident, or use of sex paraphernalia. The classic presentation is a large, tender expanding mass in the perineal or labial area. Diagnosis is by inspection and speculum exam for lacerations or expansion of the hematoma into the vagina. Treatment initially is supportive with application of a cold compress and pressure dressing. For an expanding hematoma, treatment is incision and drainage and ligation of the traumatized vessel, which may be difficult to identify as it may have retracted into the connective tissue of the perineum. The wound is then packed with gauze.

Infectious causes of acute vulvar pain include **vulvar abscess** and **acute genital herpes**. The most common vulvar mass is Bartholin's gland cyst – an abscess may form when the gland's duct is blocked. Diagnosis is based on physical exam; a normal Bartholin's gland cannot be palpated. A Bartholin's abscess is located at 5 and 7 o'clock position at the level of the vaginal introitus. Smaller abscesses (<2 cm) may resolve with conservative therapy (i.e., warm compress and sitz baths); however, incision and drainage may be warranted. Surgical drainage of the abscess and placement of a Word catheter is the treatment of choice. Leaving the Word catheter in place for 6–8 weeks ensures that the duct's outlet is well epithelialized, allowing for future drainage. For recurrent abscesses, marsupialization or excision of the gland is considered. **Vulvar abscesses** can be late infectious manifestations of skin or hair follicle infections or repaired obstetrical lacerations. Cultures should be obtained.

Genital herpes infections are STIs caused by the herpes simplex virus type 1 (HSV-1) or type 2 (HSV-2). Genital herpes infections manifest as blister lesions on the external genitalia and can also be perirectal. Primary genital herpes infections can be extremely painful with an accompanying flu-like illness; patients may require admission for intravenous antiviral therapy and pain management. Occasionally, patients may present with urinary retention. Systemic symptoms, especially pulmonary and neurologic should be addressed. Treatment with antiviral medication can shorten the duration of the primary outbreak. Secondary genital herpes infections are usually less painful and can be treated with oral antiviral medication.

Chemical irritants and the use of perfumes, exfoliating creams, and hygiene products can cause a **chemical vulvitis**. Diagnosis is usually made by history and inspection. Therapy is usually supportive. A water-based lubricant and a topical steroid ointment may be employed for severe cases.

Vagina

Acute injuries to the vagina include lacerations after sexual assault, consensual sex, or the use of sex paraphernalia. Any patient with a vaginal laceration should be screened for domestic violence, rape, or sexual abuse. The classic presentation is continuous bright red vaginal bleeding. Diagnosis is made by speculum exam. On examination, it is important to inspect the entire laceration, as it may track into the deep sulcus area of the vagina. Assessment for STIs should be performed. Repair is performed with absorbable suture. Acute painful injuries to the vagina may also be seen in the

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_80, © Springer New York 2015

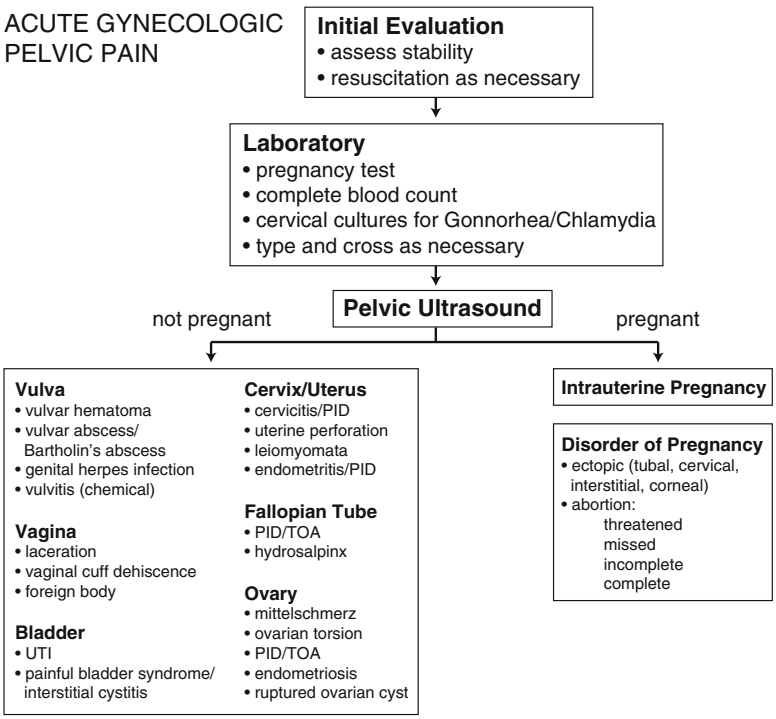

FIG. 80.1 Treatment algorithm for acute gynecological pelvic pain. *UTI* urinary tract infection, *PID* pelvic inflammatory disease, *TOA* tubo-ovarian abscess

postoperative period after a hysterectomy. Vaginal cuff dehiscence can occur when a patient engages in sexual intercourse prior to the healing of the vaginal cuff. Diagnosis is made by speculum exam. Evisceration of the bowel through the vaginal defect is occasionally noted. Treatment is closure of the cuff. The extent and mode of the repair depends on the size of the defect and the presence or absence of bowel evisceration. Small defects may be closed vaginally. Larger defects accompanied by evisceration of bowel require endoscopic repair or laparotomy.

Vaginal foreign bodies (tampons, sex paraphernalia, and other foreign bodies inadvertently left in the vaginal canal) may cause acute pelvic pain. Diagnosis is made via speculum exam and removal is performed manually or with ring forceps. Antibiotics are rarely needed, unless systemic infection or toxic shock syndrome is present.

Bladder

The most common cause of acute bladder pain is urinary tract infection (UTI). Cystitis is usually a result of growth of bacteria from the vagina and gastrointestinal tract entering into the urethra and tracking to the bladder. Clinical diagnosis is based on dysuria, urgency, frequency, and gross hematuria. Laboratory investigation includes urine analysis and culture. Treatment of bacterial UTI is with oral antibiotics. Acute injuries to the bladder can be manifestations of vaginal lacerations extending into the bladder.

Painful bladder syndrome/interstitial cystitis may cause acute bladder pain related to bladder filling and urinary urgency and frequency. This diagnosis is supported by the absence of infectious or other obvious pathology.

Cervix/Uterus

The most common cause of acute pelvic pain in the uterine corpus is miscarriage. The classic presentation is a history of a missed menses followed by vaginal bleeding and pelvic pain. The cause is a gestation that has not developed into a viable embryo, usually secondary to chromosomal abnormalities. Clinical diagnosis is made by speculum exam, pelvic exam, and pelvic ultrasound. Patients may have an open cervical os with tissue noted at the opening. The uterus is enlarged on bimanual exam and may be tender to palpation. Laboratory evaluation includes urine or serum human chorionic gonadotropin (hCG). Ultrasound examination of the pelvis is diagnostic and can delineate the location of the products and rule out other etiologies such as ectopic (fallopian tube) pregnancies. The goal of treatment is evacuation of the uterine contents. This can be done medically with the use of misoprostol or surgically by suction curettage. Patients may be candidates for observation and spontaneous completion of abortion if their blood loss is mild at presentation. Retained products of conception may develop into a septic abortion.

Uterine perforation is another cause of acute pelvic pain. This commonly occurs after a voluntary interruption of

pregnancy or other uterine surgical procedures (dilation and curettage, hysteroscopy). Imaging is performed to identify the presence of an intra-abdominal hematoma or gas in the peritoneal cavity. Though most uterine perforations go unnoticed and can be treated with observation, occasionally it is necessary to repair the defect to cease active bleeding. Laparoscopy is the gold standard for diagnosis and treatment. Broad-spectrum antibiotics should be administered if there are signs of infection. **Dysmenorrhea** is a recurrent, cyclic, crampy pelvic pain during menstrual periods. At times, this pain may be debilitating. The pain may sometimes radiate to the back and include associated symptoms such as nausea, emesis, and headaches. Dysmenorrhea is caused by an excess of prostaglandin production. Pregnancy must be ruled out. Pelvic ultrasound is utilized to help rule out other anatomic causes. Nonpharmacologic intervention can be utilized; heat, dietary changes, and vitamin and herbal treatment have variable success. Treatment is usually accomplished with nonsteroidal anti-inflammatory drugs (NSAIDs) and oral contraceptive pills.

Leiomyomata, commonly known as fibroids, are benign uterine tumors and can occasionally be a cause of acute pelvic pain and heavy, abnormal vaginal bleeding. As fibroids expand, they can outgrow their blood supply leading to necrosis and subsequent intense pelvic pain. Clinical diagnosis is made by pelvic exam and ultrasound to confirm uterine leiomyomata. Immediate pain concerns can be alleviated with NSAIDs, narcotics, and hydration. Ultimately, the goal of treatment is to remove the offending pathology. In reproductive-age women desiring future childbearing, or women desiring retention of her uterus, myomectomy is performed. Otherwise, hysterectomy is indicated.

Endometritis is an infectious process of the uterine lining and may occur after vaginal delivery or cesarean section. Endometritis can also present outside of pregnancy, as part of the continuum of pelvic inflammatory disease/tubo-ovarian abscess (discussed later). Treatment is accomplished usually with oral antibiotic therapy; parenteral antibiotics are reserved for systemic infections or failed oral antibiotic therapy.

Fallopian Tube

The most common cause of acute pain of the fallopian tube in reproductive-aged women is **ectopic/tubal pregnancy**. Patients may have a past history of pelvic infections, especially STIs, multiple sexual partners, previous ectopic pregnancy, and previous tubal surgery or tubal sterilization. A patient with an intrauterine device who presents with a positive pregnancy test is at risk for ectopic pregnancy. The classic signs and symptoms are pelvic pain, amenorrhea, and vaginal bleeding 6–8 weeks after a normal menstrual period. Sites other than the tube that can have an ectopic pregnancy include the cervix, ovary, and interstitial or corneal portion of the uterus. Clinical diagnosis is made by serum human chorionic gonadotropin (hCG) and pelvic ultrasound. Vital signs may reveal orthostatic hypotension and tachycardia secondary to hemorrhage. The goal of treatment is the removal of the gestation. This can be accomplished medically with methotrexate in hemodynamically stable patients. Surgical management involves the removal of the gestational sac without removal of the fallopian tube (salpingostomy) or removal of the fallopian tube with the gestational sac (salpingectomy). Most surgical cases can be done laparoscopically with laparotomy reserved for unstable patients.

Ovary

Common causes of acute pelvic pain of the ovary include: mittelschmerz (pain with ovulation), ovarian neoplasm with or without concomitant torsion, pelvic inflammatory disease/tubo-ovarian abscess (PID/TOA), and endometriosis.

Mittelschmerz is a mid-cycle, unilateral pelvic pain secondary to the release of an ovarian follicle. Pain may last for 2–3 days and is usually self-limited. NSAIDs can be employed as first-line therapy.

Ovarian torsion is a surgical emergency and classically presents as a sudden sharp pelvic pain. It is the result of the ovary twisting on its blood supply. The presence of an ovarian neoplasm may predispose to torsion. Pain is usually unilateral. The causes of ovarian torsion are unknown, but are associated with adnexal masses (ovarian or fallopian tube origin) usually 5 cm or larger. Clinical diagnosis is made by pelvic examination and by ultrasound assessing for the presence or absence of an ovarian mass; blood flow to the ovary can be measured at this time. The treatment is surgical decompression of the ovarian torsion, and the removal of adnexal mass (if present) may result in resolution and avoidance of necrosis. Evidence of necrosis may warrant removal of the affected adnexa.

Pelvic inflammatory disease/tubo-ovarian abscess (PID/TOA) is a spectrum of infectious processes commonly occurring in reproductive-age, sexually active women. The cause is related to bacterial infection of the genital tract, usually by gonorrhea and chlamydia; however, infections are often multi-bacterial. Purulent vaginal discharge may be present. Clinical diagnosis is made by pelvic exam. The presence of cervical motion tenderness, uterine tenderness, and bilateral adnexal tenderness in the presence of a negative pregnancy test confirms the diagnosis of PID. The diagnosis of TOA requires ultrasound. Both oral and intravenous antibiotics may be used, depending upon the severity of the disease, size of the abscess, and the reliability of the patient. Surgical treatment includes drainage of the abscess either by interventional radiology or laparoscopy. Removal of the infected adnexa and uterus is reserved for larger abscesses 6 cm or greater or failed antibiotic therapy.

Endometriosis is another cause of cyclic or continuous pelvic pain. Endometriosis is the presence of endometrial

glands and stroma outside the endometrial cavity. It can occur anywhere in the pelvis and is frequently seen on the ovary where it may manifest as small implants or large ovarian masses called endometriomas. Distal endometriotic implantations (diaphragm, lung, and brain) have been reported. The cause of endometriosis is unclear; many theories exist. The most widely accepted theory is coelomic metaplasia. Diagnosis is made by laparoscopy and biopsy of the implants to confirm the disease process, although its appearance is variable and can be difficult to distinguish from nascent peritoneal tissue. Larger endometriomas can present as ovarian neoplasms. The goal of treatment is primarily pain control and halting progression of the disease process. Oral contraceptive pills may help with acute periods of pain. Also, gonadotropin-releasing hormone (GnRH) agonists (e.g., leuprolide) can be used for temporary relief. Both oral contraceptives and GnRH agonist therapy treat the symptoms of the disease process. Regression of lesions can occur with GnRH agonists but will recur once therapy is discontinued. Definitive treatment is surgical with total abdominal hysterectomy and bilateral oophorectomy along with resection of all visible endometriosis implants. Scarring and obliteration of normal tissue planes should be anticipated since endometriosis can invade adjacent pelvic organs.

A **ruptured ovarian cyst** is an ovarian cause of acute pelvic pain. The rupture of an ovarian cyst or a corpus luteum can occur after physical activity, sexual intercourse, or de novo, resulting in generalized acute pelvic pain. Most cases can be observed with supportive measures such as fluids and pain medication. A ruptured corpus luteum with massive hemorrhage necessitating blood products or surgical intervention is uncommon.

81
Evaluation and Management of Adnexal Masses

Alfred Guirguis and Elizabeth Weldon

Introduction

The adnexa refers to the region that usually contains the ovary, fallopian tube, any associated vessels, ligaments, and connective tissue. At times, pathology coming from the uterus, bowel, retroperitoneum, and/or metastatic disease may also be within the adnexal region. Although variable, the prevalence of adnexal masses is highest during premenopausal years at approximately 6–8 % and decreases in the menopausal state.

Of utmost concern with these adnexal masses is to rule out malignancy. Approximately 35 % of all complex adnexal masses in adolescent girls under the age of 18 are malignant and most of those are germ cell tumors. In the premenopausal female with an adnexal mass, the overall risk of malignancy ranges between 6 % and 11 %, whereas in a postmenopausal patient it ranges from 29 % to 35 %.

Choosing the best radiographic modality for initial evaluation of a pelvic mass can be challenging. Ultrasound (US) has been shown to be the most valuable diagnostic study since it does not emit radiation, does not require a potentially nephrotoxic dye load, and has a sensitivity and specificity of 86–91 % and 68–83 %, respectively. In addition to the ultrasound features, the specific location of the adnexal mass is crucial to know because a retroperitoneal mass has severely different consequences than an intraperitoneal mass, and location can dictate which surgical specialty to consult.

The following three tables define terminology pertaining to the management of adnexal masses (Table 81.1), describe potential ultrasound features (Table 81.2), and list the differential diagnoses (Table 81.3).

Once an intraperitoneal mass is seen on ultrasound, a BHCG is obtained to dictate the appropriate treatment algorithm as depicted in Fig. 81.1.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_81, © Springer New York 2015

TABLE 81.1. Terminology for treatment of adnexal masses.

	Definitions
Surveillance	Serial pelvic exams and transvaginal ultrasound, initially every 6–8 weeks
Surgery	Depending on the etiology and the size of the adnexal mass, this can be performed either by laparoscopy or laparotomy
Ectopic pregnancy	Suspect in women with abdominal pain/vaginal bleeding with beta HCG >1,500 without intrauterine pregnancy, or if ectopic pregnancy visualized on transvaginal US. This can be managed medically with methotrexate or surgically
Ovarian torsion	Refers to the twisting of the ovary/fallopian tube on its ligamentous pelvic attachments, which frequently leads to a compromise of the blood supply
Tubo-ovarian complex (TOA)	Inflammation of the adnexal structures causing an agglutination of the fallopian tube and ovary. Managed with broad-spectrum antibiotics +/− surgical drainage
Endometrioma	Cyst comprised of ectopic endometrial tissue in women with endometriosis
Follicular cyst	Simple cyst that occurs when a mature follicle fails to ovulate
Corpus luteum cyst	Occurs when the corpus luteum fails to involute, or in order to sustain a pregnancy during the first trimester
Hydrosalpinx	Cystic dilation of the fallopian tube due to distal occlusion, usually from previous infection
Leiomyoma	Benign smooth muscle tumor of the uterus
Hemoperitoneum	Blood in the peritoneum (from ruptured ectopic or hemorrhagic cyst) that can lead to irritation and symptoms of peritonitis
Tumor markers	
CA 125	Marker for epithelial ovarian cancer
OVA-1	Marker for epithelial ovarian cancer
AFP	Marker for endodermal sinus tumor
hCG	Marker for choriocarcinoma, embryonal tumor
LDH	Marker for dysgerminoma, endodermal sinus tumor
Inhibin	Marker for granulosa cell tumor
Androgens	Marker for Sertoli-Leydig and granulosa cell tumors

TABLE 81.2. Ultrasound features of adnexal masses.

Low risk features	High risk features
Simple	Complex
Smooth walls	Nodular/papillary
Thin walls, no septations	Thick septations
No ascites	Present ascites
<10 cm in size	>10 cm in size

TABLE 81.3. Differential diagnosis of adnexal mass.

Adnexal mass		
Gynecologic	Benign	Simple Follicular cyst Hemorrhagic cyst Corpus luteum cyst Paratubal cyst Hydrosalpinx
		Complex Endometrioma Tubo-ovarian complex Ovarian torsion Ectopic pregnancy
	Tumors	Benign Mature teratoma (dermoid) Serous/mucinous cystadenoma Brenner's tumor
		Malignant Epithelial carcinoma Sex cord stromal tumor Germ cell tumor
Non-gynecologic	Benign Diverticular/appendiceal abscess Pelvic kidney Ureteral/bladder diverticulum	
	Malignant GI cancer Retroperitoneal sarcoma Lymphoma Metastases	

ADNEXAL MASSES

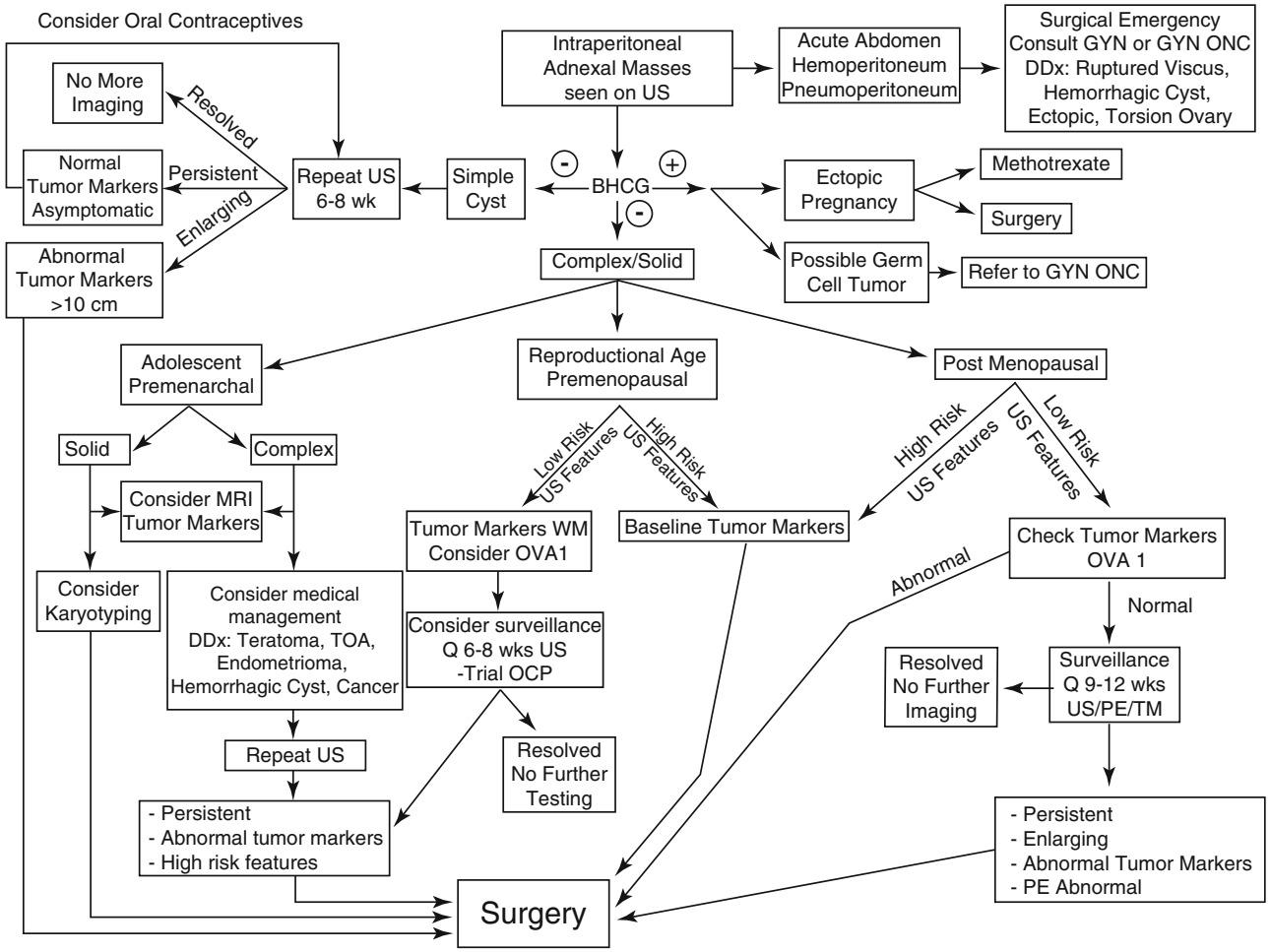

FIG. 81.1 Treatment algorithm for adnexal masses. *OCP* oral contraceptives, *US* ultrasound, *DDx* differential diagnosis, *Gyn* gynecologist, *Gyn Onc* gynecologic oncology, *TOA* tubo-ovarian complex, *PE* physical exam

82
Pelvic Inflammatory Diseases

Jaqueline Blank

Pathogenesis

Pelvic inflammatory disease (PID) encompasses infections of the uterus (endometritis or myometritis), fallopian tubes (salpingitis), ovaries (oophoritis), broad ligaments (parametritis), and pelvic peritoneum (peritonitis). PID accounts for up to 2.5 million outpatient visits and 350,000 emergency department visits per year for women. *It is the leading cause for hospital admissions among women of reproductive age*, though its incidence is decreasing due to more aggressive screening for chlamydial infections.

PID is a polymicrobial disorder that results from disruption of the normal bacterial flora of the vagina and cervix, including *Streptococcus agalactiae*, gram-negative cocci, and anaerobes (Fig. 82.1). The two initiating pathogens are *Neisseria gonorrhoeae* and *Chlamydia trachomatis*. PID initiated by *N. gonorrhoeae* tends to be more acute and severe than that caused by *C. trachomatis*. By definition, this is a sexually transmitted infection, not one caused by medical procedures or pregnancy. Approximately 50 % of males may be asymptomatic at the time of transmission of infection.

Risk factors for PID include young age at first sexual intercourse, multiple sexual partners (which represents a 3.4-fold increase in the risk of PID if a woman has had four or more sexual partners within 6 months), and a history of sexually transmitted infections. Barrier contraception is protective against PID. Patients with PID commonly present with acute or subacute lower abdominal pain, especially with coitus, during or after their menses. In the 7 days preceding menses, cervical mucus is thinnest and most susceptible to penetration by pathogens. Additional clinical criteria for PID include cervical motion tenderness, adnexal or uterine tenderness, abnormal uterine bleeding, purulent vaginal discharge, fevers, and chills.

Diagnosis

Minimal criteria for the diagnosis of PID include abdominopelvic pain in a sexually active woman plus cervical, uterine, or adnexal tenderness. Criteria that increase the specificity of the diagnosis include temperature greater than 38.3 °C, white blood cells seen on saline preparation of vaginal secretions, cervical exudates, and cervical friability. Laboratory evaluation that supports the clinical diagnosis of PID includes elevated C-reactive protein or sedimentation rate and positive cervical cultures. If a diagnosis of PID is confirmed, these patients should be offered testing for human immunodeficiency virus (HIV).

Differential Diagnosis

The differential diagnosis includes appendicitis, diverticulitis, inflammatory bowel disease (IBD), nephrolithiasis, cystitis, pyelonephritis, ectopic pregnancy, ovarian torsion, ruptured ovarian cyst, or degenerating fibroids.

Triage

Once the diagnosis is established, it is important to determine whether a patient needs to be admitted for intravenous antibiotic therapy. Generally, patients who require hospitalization included pregnant patients, severely ill patients (with nausea, vomiting, and fever), patients unable to tolerate oral antibiotics, patients who may be noncompliant with oral antibiotics, patients with suspected tubo-ovarian abscess, patients in whom the diagnosis is uncertain, and patients who have not responded to oral antibiotics in the outpatient setting.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_82, © Springer New York 2015

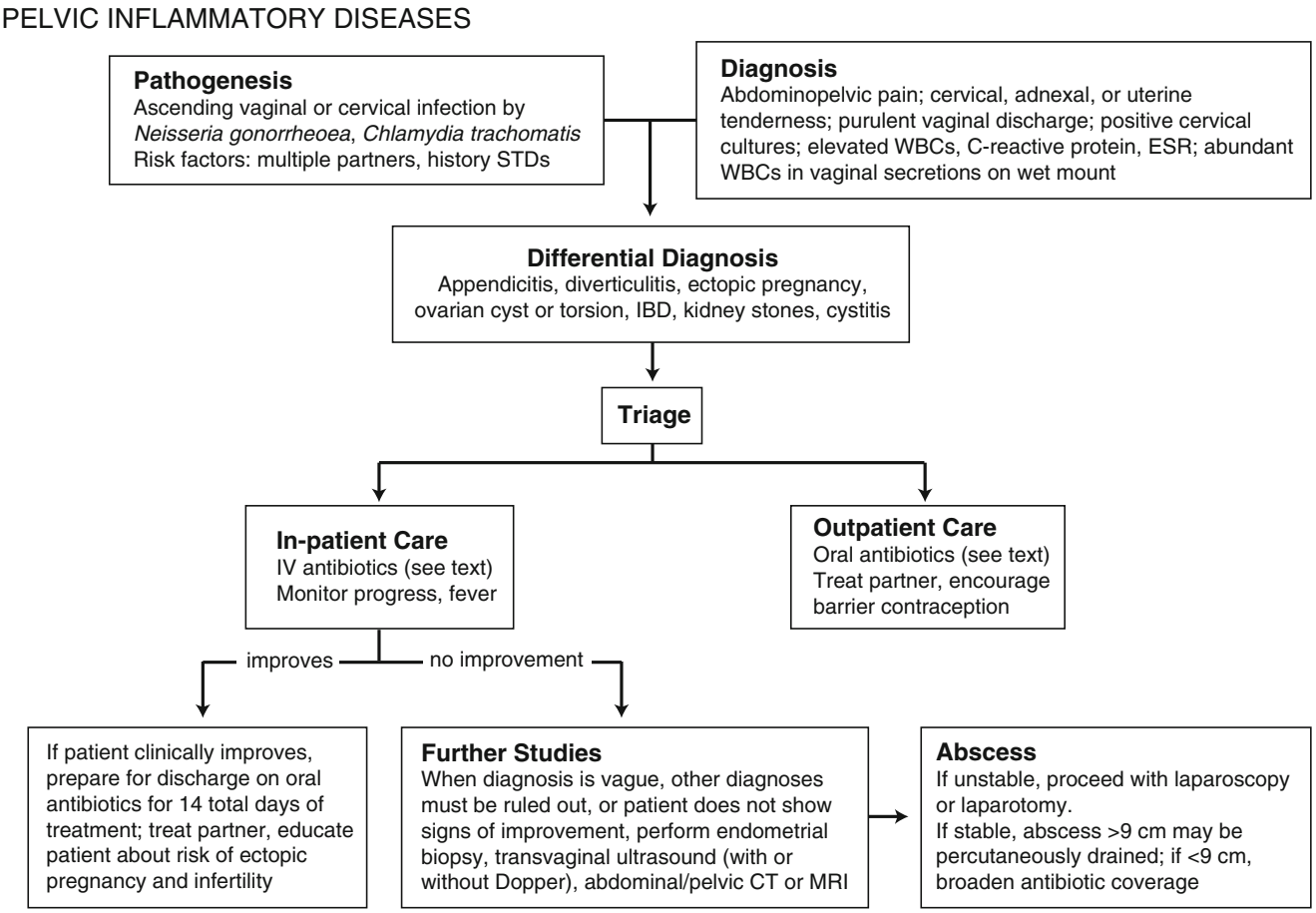

FIG. 82.1 Treatment algorithm for pelvic inflammatory diseases. *STDs* sexually transmitted diseases, *WBCs* white blood cells, *ESR* erythrocyte sedimentation rate, *IBD* inflammatory bowel disease, *IV* intravenous, *CT* computed tomography, *MRI* magnetic resonance imaging

Inpatient Care

Those admitted for intravenous antibiotic therapy can be treated with one of the following regimens:

(a) Cefotetan 2 g IV q 12 h or cefoxitin 2 g IC q 6 h *plus* doxycycline 100 mg IV q I2h
(b) Clindamycin 900 mg IV q 8 h *plus* either gentamicin IV/IM load of 2 mg/kg body weight then 1.5 mg/kg q 8 h *or* gentamicin 3–5 mg/kg IV/IM once daily
(c) Ampicillin/sulbactam 3 g IV q 6 h *plus* doxycycline 100 mg IV q 12 h

Only regimen (b) can be used in pregnant patients. All other regimens contain antibiotics that are contraindicated in pregnancy.

Patients are continued on parenteral antibiotics for 24 h after the improvement of symptoms. If these criteria are met, patients can be discharged on one of the following regimens of oral antibiotic therapy:

(a) Doxycycline 100 mg twice a day
(b) Clindamycin 450 mg four times a day

Regimen (b) is preferable for patients in whom a tubo-ovarian abscess is suspected. Patients must be treated for a total of 14 days. Treatment of the sexual partner and use of barrier contraception should be encouraged.

These patients should be reevaluated within a week. If they continue to show improvement, they should complete their antibiotic course. Upon completion they can be followed-up routinely.

Outpatient Care

Patients who met criteria for outpatient therapy upon initial evaluation can be treated with one of the following regimens:

(a) Ceftriaxone 250 mg IM once *plus* doxycycline 100 mg PO twice a day for 14 days *with* or *without* metronidazole 500 mg PO twice a day for 14 days

(b) Cefoxitin 2 g IM once *plus* probenecid 1 g PO concurrently *plus* doxycycline 100 mg PO twice a day for 14 days *with* or *without* metronidazole 500 mg PO twice a day for 14 days
(c) Other 3rd-generation cephalosporin (ceftizoxime, cefotaxime) *plus* doxycycline 100 mg PO twice a day for 14 days *with* or *without* metronidazole 500 mg PO twice a day for 14 days

Cefoxitin provides better anaerobic coverage; ceftriaxone provides better coverage for N. gonorrhoeae. *Metronidazole provides better coverage for bacterial vaginosis, which is frequently associated with PID. Treatment of the sexual partner and use of barrier contraception should be encouraged.* For patients who do not improve, further studies should be done to rule out an abscess.

Further Studies

If the patient is not improving on IV antibiotics, if an adnexal mass is palpable, or if the patient is too tender for a proper pelvic exam, further studies are warranted. Endometrial biopsy may lend histopathologic proof of endometritis. Imaging studies include transvaginal ultrasound (TVUS) with or without Doppler, MRI, and abdominal CT. A TVUS may show thickened or filled fallopian tubes with or without free pelvic fluid. A CT or MRI may show evidence of a pelvic or tubo-ovarian abscess.

Abscess

When imaging studies reveal the presence of an abscess, one of the following management protocols can be pursued:

(a) If the patient becomes hypotensive, tachycardic, and acidotic or manifests peritoneal signs, or if there is evidence of sepsis or abscess while on antibiotics, a laparoscopy or laparotomy is warranted to remove the abscess. These patients should be counseled preoperatively about the possibility of a hysterectomy and/or possible removal of the fallopian tubes and ovaries.
(b) If the patient is not worsening nor improving on antibiotics, image-guided drainage of the abscess is warranted. The patient should continue IV antibiotics with additional anaerobic coverage.
(c) If the patient is hemodynamically stable, the abscess is less than 9 cm, the patient has had adequate response to antibiotics, and the patient is premenopausal, then the patient should receive broad-spectrum antibiotics. If there is no significant improvement in 24–48 h, these patients should undergo surgical assessment.

Once the diagnosis of PID is made, patients should be counseled about the following sequelae of the disease:

- Women with PID have an increased infertility rate. The degree of infertility appears to be directly proportional to the number of episodes of PID. Women with three or more episodes have infertility rates up to 60 %.

Part X
Critical Care

83
Postoperative Glucose Management

Jennifer Leung Schoenberger and David Baldwin Jr.

Introduction

Uncontrolled hyperglycemia is a risk factor for postoperative wound infections. Most current guidelines recommend a target blood glucose of 150 mg/dl in hospitalized patients with hyperglycemia. The risk for infection and for an increased length of stay rises with blood glucose > 180 mg/dl, while length of stay and the risk of mortality rise with blood glucose < 80 mg/dl. Therefore, it is important to have protocols in place that are able to maintain most blood glucose levels between 100 and 180 mg/dl while avoiding hypoglycemia.

Target Population

Most patients admitted with a previous history of type 2 diabetes will require continued inpatient therapy whether they were previously treated with oral agents or insulin. All patients with type 1 diabetes require uninterrupted treatment with at least their current doses of basal insulin (NPH or glargine) in order to avoid developing diabetic ketoacidosis. Other patients, not previously diagnosed with type 2 diabetes, may develop hyperglycemia after major surgery. This may be a temporary reaction to the stress of surgery or may reflect previously undiagnosed diabetes. Other risks for developing postoperative hyperglycemia include total parenteral nutrition, enteral tube feeds, and treatment with glucocorticoids (Fig. 83.1).

Initial Approach

Blood glucose testing using point of care meters should be initiated every 6 h for all patients with a history of diabetes and all patients who develop a serum glucose > 150 mg/dl on routine laboratory testing. Oral antidiabetic agents such as metformin or the sulfonylureas are best discontinued during hospitalization due to their lack of efficacy or their risk for causing hypoglycemia. HgbA1C should be measured in all patients with a history of diabetes and those with new-onset hyperglycemia in order to assess blood glucose control for the past 3 months. Type 2 diabetes is diagnosed by HgbA1C > 6.5 %. This will allow differentiation of prior undiagnosed diabetes from stress-induced hyperglycemia and will guide diabetes treatment initiation or modification upon hospital discharge.

Insulin Therapy for Intensive Care Patients

An intravenous (IV) insulin infusion protocol should be started on all postoperative patients in the intensive care unit (ICU) with blood glucose > 160–180 mg/dl. Blood glucose is measured hourly. The rate of IV insulin is titrated hourly by the ICU nursing staff according to the protocol to maintain blood glucose within a target range of 140–160 mg/dl. The protocol should also include components for the prevention and treatment of hypoglycemia as well as for transition to subcutaneous insulin when the patient is ready for transfer to a general surgical unit. All patients with type 1 diabetes and all patients with type 2 diabetes who are requiring ≥ 1 unit IV insulin/hour should be transitioned to once daily subcutaneous basal insulin glargine. To calculate the first dose of insulin glargine, multiply the current stable IV insulin unit/hour rate by 20. Continue the IV insulin infusion for 3 h after the first dose of glargine. Continue dosing insulin glargine every 24 h and adjust the dose daily to maintain the 6 AM blood glucose in the 100–120 mg/dl range. Once a patient is eating, rapid-acting insulin analog (lispro, aspart, or glulisine) is added with each meal beginning with one-sixth of the insulin glargine dose. Rapid-acting mealtime insulin is adjusted daily to maintain 12 PM, 6 PM, and 11 PM blood glucose levels in the 140–180 mg/dl range.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_83, © Springer New York 2015

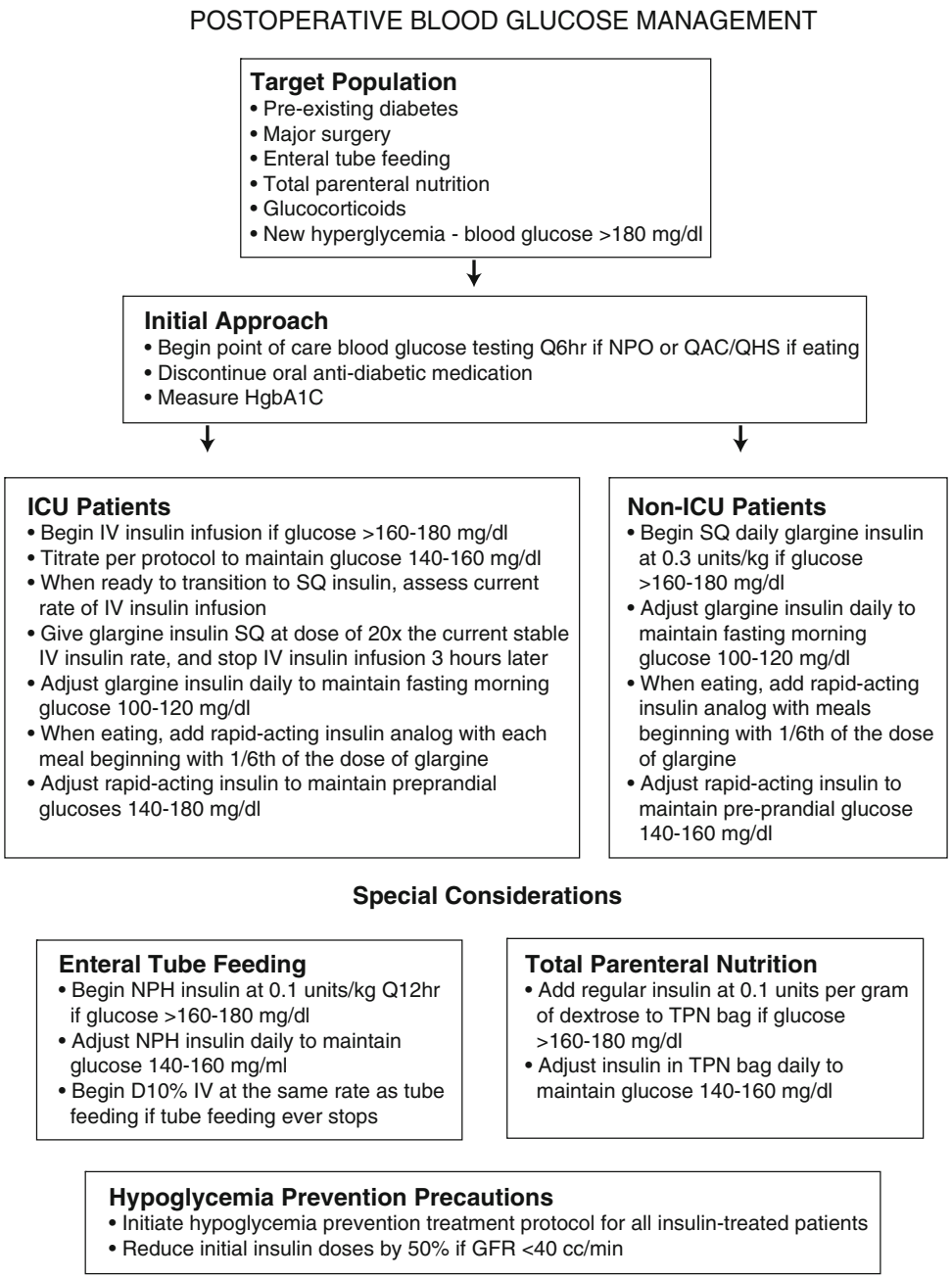

FIG. 83.1 Algorithm for postoperative blood glucose management. *NPO* nil per os, *QAC* before every meal, *QHS* every bedtime, *SQ* subcutaneous, *NPH* neutral protamine Hagedorn, *IV* intravenous, *TPN* total parenteral nutrition, *GFR* glomerular filtration rate

Insulin Therapy for the General Surgical Floor

Start glargine insulin at 0.3 units/kg/day on non-ICU patients with blood glucose > 160 mg/dl following surgery. Since glargine is non-peaking, it is ideal for NPO patients who only require basal control of hyperglycemia. Adjust daily to maintain the 6 AM blood glucose 100–120 mg/dl. Patients who take NPH insulin or the NPH-based premix insulins 70/30 or 75/25 BID can be converted to once daily glargine by giving 50 % of the total daily dose of NPH/NPH-based premix as glargine. Once a patient begins to eat, rapid-acting insulin can be dosed with meals beginning with one-sixth of the insulin glargine dose per meal and titrated daily, aiming for blood glucose 140–160 mg/dl. Decrease insulin doses by 20 % if blood glucose < 100 mg/dl and increase by 20 %

if blood glucose > 200 mg/dl. If glargine insulin is being started on a hospitalized patient with chronic renal failure (estimated glomerular filtration rate (GFR) < 40 cc/min), it is recommended to reduce the initial dose of glargine by 50 % because of the increased risk for hypoglycemia attendant with diminished renal clearance of insulin.

Enteral Tube Feeding

Enteral tube feeding is often needed to support patients after major surgery. Even if a history of diabetes is absent and if blood glucose levels were normal prior to tube feeds, it is common for enteral tube feeds to provoke hyperglycemia in these patients, especially when elderly. Blood glucose > 160–180 mg/dl should be treated with basal insulin. We recommend giving NPH insulin every 12 h beginning with 0.1 units/kg/dose. NPH has the advantage of allowing dose titration every 12 h, in order to maintain blood glucose in the 140–160 mg/dl range. Tube feedings in the hospital often stop abruptly because of tube loss or malfunction, imaging, or procedures. In order to prevent hypoglycemia in this situation, immediately start a 10 % IV dextrose infusion at the same rate as the tube feeding, and continue until the last dose of NPH has worn off or until feedings are resumed. Often tube feeds may only be administered overnight for 12 h. In this situation, a single dose of NPH insulin given at the start of feeding usually is the only insulin required for 24 h.

Total Parenteral Nutrition

After surgery, especially open abdominal procedures, it is common to develop an ileus that will delay intake of oral nutrition or tube feeds. These patients are often started on total parenteral nutrition (TPN). Like patients who receive enteral tube feeds, TPN patients commonly develop hyperglycemia with or without a prior history of diabetes. If blood glucose is > 160–180 mg/dl, the safest and most effective way to give insulin is by adding regular insulin directly to the bag of TPN beginning with 0.1 units of insulin per gram of dextrose. The dose is adjusted daily to maintain a blood glucose target of 140–160 mg/dl.

Hypoglycemia Prevention and Treatment

Hospitalized patients receiving insulin are always at risk for hypoglycemia due to renal or hepatic impairment, sepsis, and erratic delivery of nutritional support. All patients receiving insulin should have a hypoglycemia prevention/treatment protocol in place so that if a patient develops a blood glucose < 70 mg/dl, orders for treatment are not delayed. Fifteen to twenty grams of carbohydrate (juice, glucose tablets or gel) is effective if they are able to take PO, or 50 ml of 50 % dextrose IV if they are NPO. Blood glucose should be rechecked every 20 min until normal and the patient's insulin regimen re-titrated as needed.

84
Oliguria

Elizabeth A. Hooper

Definition

Oliguria is the sudden decrease in urine output – most often defined as <400 mL/day or <0.5 mL/kg/h (Fig. 84.1). It may be secondary to a multitude of factors. At the nephron level, acute oliguria occurs in the setting of a decreased glomerular filtration rate (GFR). With a drop in GFR, the kidney signals for sodium and water retention, leading to resultant hyperkalemia and acidosis. Additionally decreased excretion of creatinine and blood urea nitrogen occurs due to the lower filtration rate.

When oliguria progresses to acute renal failure (ARF), the postsurgical patient has an approximate 50 % mortality rate, most commonly from sepsis or multiorgan failure. Oliguria has progressed to ARF when the glomerular filtration rate drops from greater than 100 mL/min to less than 30 mL/min.

Classification of Oliguria

Three classifications make up the types of underlying causes of oliguria: prerenal, intrinsic, and postrenal (or obstructive uropathy). Prerenal and postrenal etiologies should be investigated and excluded first because they are often reversible if treated in a timely manner. Initial management entails a detailed history, physical exam, and blood and urine studies, which should be sent prior to instituting therapy.

Prerenal Causes

Prerenal ARF is characterized by diminished renal blood flow and accounts for the majority (60–70 %) of renal failure cases. Prerenal renal failure may be the precursor to intrinsic renal failure from ischemia-induced acute tubular necrosis (ATN). Assessment of volume status, hemodynamics, and medications may reveal potential prerenal causes of oliguria.

Laboratory findings demonstrate a fractional excretion of sodium (FENa) less than 1 %, urine osmolality greater than 500 mOsm, and urinary sodium less than 20 mmol/L. These values are consistent with functional kidneys attempting to conserve intravascular volume through reabsorption of sodium and water.

The most likely cause of prerenal ARF is due to malperfusion of the renal vasculature secondary to hypovolemic shock. This may be due to a deficit of crystalloid hydration or secondary to hemorrhage. Alternatively, it may be a result of sepsis causing systemic vasodilation and a functional appearance of hypovolemia.

Initial evaluation of volume status is through history and physical exam. Signs of hypovolemia include sinus tachycardia, orthostatic and/or supine hypotension, and mucocutaneous changes (decreased skin turgor, delayed capillary refill). Mental status changes including dizziness and decreased wakefulness may occur. A drop in the urine output should always be a potential sign of hypovolemia.

The patient may have a loss of intravascular volume secondary to hemorrhage, gastrointestinal losses (vomiting, diarrhea, or nasogastric suctioning), fluid shifts from pancreatitis, small bowel or large bowel obstruction, or ascites. Renal losses may drop the intravascular volume in patients with diabetic ketoacidosis (DKA) or nonketotic hyperosmolar states where inappropriate diuresis occurs. Insensible losses from burns, fevers, or open abdomens are also potential sources of hypovolemia.

In all of these scenarios, the patient has a suboptimal preload volume and a resulting decrease in the cardiac output (CO). The underlying cause must be identified and treated in order to return the volume status to normal. Isotonic fluid replacement should be given as a fluid bolus. A hematocrit or hemoglobin should be obtained and transfusion of packed red blood cells should be transfused when indicated. Volume and rate of crystalloid or colloid (blood transfusion) will vary depending on the severity of the hypovolemia and must be balanced against the patient's overall status. Pulmonary edema, hypoxia, and cardiac compromise due to volume overload are all potentional complications. The abnormal vital signs seen in the workup of hypovolemic, prerenal

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_84, © Springer New York 2015

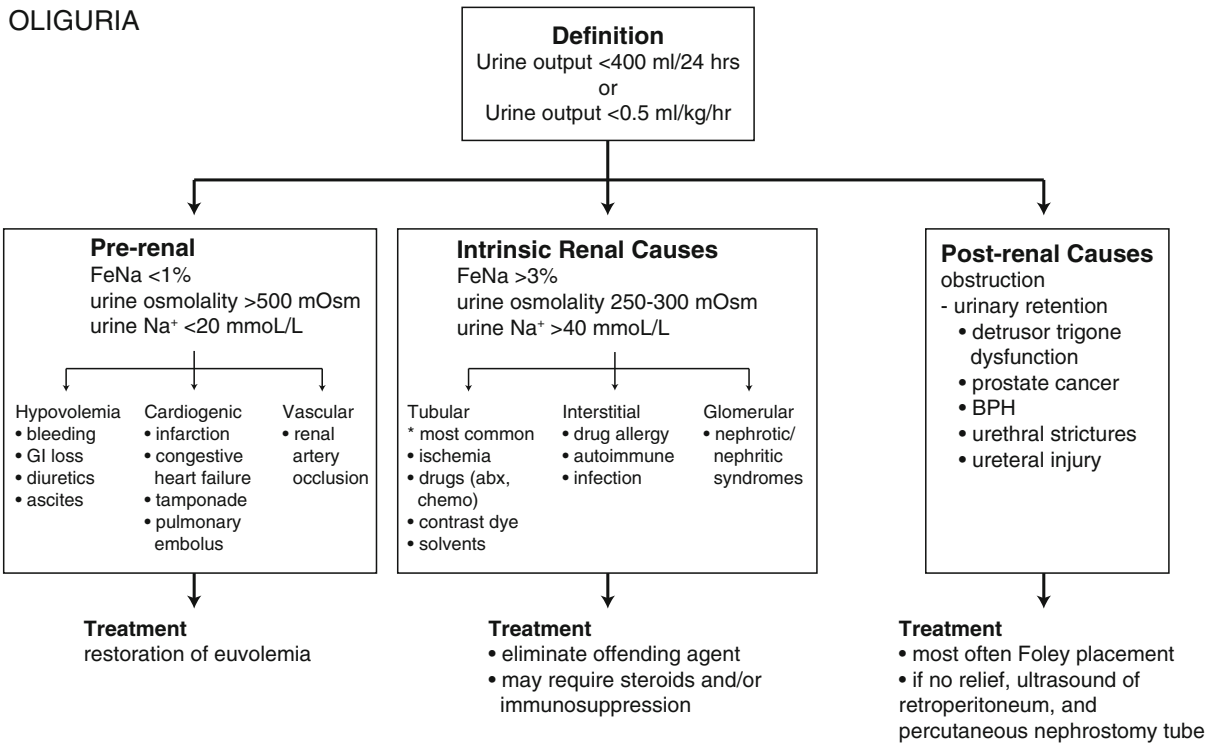

FIG. 84.1 Algorithm for treatment of oliguria. *FeNa* fractional excretion of sodium, *GI* gastrointestinal, *BPH* benign prostatic hypertrophy, *US* ultrasound

oliguria should resolve with fluid. Occasionally when multiple fluid boluses are required, a central venous line may assist in estimating the venous return to the right heart. In extremely ill patients with severe cardiac or lung dysfunction, a Swan-Ganz catheter may provide valuable information for hydration endpoints via measurement of central venous pressure, pulmonary capillary wedge pressure, and cardiac output. Patients in septic shock may require invasive monitoring and addition of vasopressor support once the preload has been restored.

Cardiac dysfunction is another source of prerenal failure. Decreased renal perfusion may occur in the presence of a myocardial infarction, significant pulmonary embolus, congestive heart failure, or cardiac tamponade. In these scenarios patients have low urine output but symptoms of fluid overload, including complaints of chest pain or shortness of breath along with jugular venous distension, rales on auscultation, atrial fibrillation, presence of a new murmur, and/or wheezing.

Treatment for cardiac causes is centered around improving the patient's cardiac output. In addition to restoring preload, addition of inotropes may be necessary. Macrovascular causes can lead to decreased renal profusion in the setting of adequate volume and cardiac output. Examples include renal artery or vein occlusion secondary to thrombus, thromboembolism, arterial stenosis secondary to atherosclerosis, or a dissecting aneurysm. Postoperative transplant recipients are at high risk for these macrovascular causes, and their presence should be quickly ruled out with a duplex ultrasound of the graft kidney.

Many medications can directly affect renal physiology. The most common are angiotensin-converting enzyme (ACE) inhibitors and nonsteroidal anti-inflammatory drugs (NSAIDS). Both types of drugs interrupt the autoregulation of the kidney. ACE inhibitors cause efferent dilation of arterioles as they leave the juxtaglomerular apparatus and create a relative low flow through the nephrons. NSAIDs inhibit prostaglandin synthesis, leading to vasoconstriction of the afferent arterioles. The effect is low flow through the juxtaglomerular apparatus leading to sodium and water retention. The cornerstone of treatment is discontinuation of the offending medication.

Renal Causes

Renal or intrinsic acute renal failure occurs when there is damage to the renal parenchyma and accounts for approximately 25–40 % of the causes of oliguria.

Laboratory studies typically reveal a FENa greater than 3 %, urine osmolality between 250 and 300 mOsm, and urine sodium greater than 40 mmol/L. Parenchymal damage leads to an inability to concentrate the urine appropriately. Therefore, despite potential low-flow states, the kidney still loses

inappropriate amounts of sodium and water. More sodium is lost in the urine than appropriate. Intrinsic renal failure is divided into subgroups based on the location of the failing parenchyma: tubular, interstitial, glomerular, and vascular.

Tubular Causes

Acute tubular necrosis (ATN) accounts for the majority of intrinsic renal causes of oliguria and/or acute renal failure. The most likely inciting event of ATN is ischemia followed by toxins. Ischemia is often due to extension of prerenal causes of oliguria progressing to tubular damage. Once ischemia begins, the GFR does not improve even with improvement in preload. Common toxins that lead to ATN include antibiotics (aminoglycosides or vancomycin) or radiocontrast dye. The risk of renal failure is proportional to the volume of contrast infused. Conventional angiograms carry the highest risk of contrast-induced ATN. Patients with preexisting renal dysfunction, diabetes, hypertension, congestive heart failure, or shock are at higher risk than the healthy population. Other less common agents causing ATN include chemotherapeutics (cisplatin) immunosuppressants (tacrolimus, cyclosporine), antifungals (amphotericin), heme pigments (myoglobin, hemoglobin), solvents (ethylene glycol), or light chains from myeloma.

Treatment is mainly supportive and includes reversal of ischemia, removal of toxins, and maintenance of euvolemia. Management includes adequate hydration, maintaining CO, and avoiding vasoconstriction. *N*-acetyl-cytsteine has been used in many trials as a free radical scavenger; currently the benefit is equivocal, but the medication is often used with adequate hydration in an attempt to prevent ATN in high-risk patients who require contrast studies.

Interstitial Causes

The most common causes are acute interstitial nephritis and glomerulonephritis. Acute interstitial nephritis usually presents with oliguria, fever, rash, eosinophilia, and occasionally eosinophiluria. This usually occurs in response to an allergic drug reaction and an autoimmune disease or in the setting of an infection or infiltrating disease of the kidney.

Treatment for interstitial causes of oliguric ATN includes elimination of any possible offending agents and biopsy of the kidney, and, in some cases, immunosuppression with corticosteroids and/or plasmapheresis may be required.

Glomerular Causes

Glomerular diseases include focal segmental glomerular sclerosis, and IgA nephropathy and subsequent renal failure are secondary to inflammation and/or occlusion of the glomerular tuft. At the microscopic level, glomerular capillary thrombosis or occlusion occurs. Patients may present with hypertension, proteinuria, hematuria, and thrombocytopenia, and workup may show nephritic or nephritic syndrome. Treatment is similar to that for interstitial renal diseases.

Postrenal Causes

Most cases of postrenal oliguria are secondary to urinary retention. A common cause of urinary retention is due to discoordination of the detrusor and trigone muscles following pelvic surgery, hernia repair, or anorectal procedures. Other common causes include benign prostatic hypertrophy and ureteral strictures. Less frequent etiologies of post-obstructive uropathy include cervical cancer, prostate cancer, fibrosis of the retroperitoneum, pelvic mass, neurogenic bladder, or ureteral stricture. Iatrogenic injury should also be included in the differential diagnosis in patients undergoing intra-abdominal or pelvic operations.

The bladder volume should be established in the workup of a postrenal source of oliguria. On physical exam, the patient may complain of suprapubic fullness with an inability to void. Many hospitals have bladder scans that provide an ultrasound estimate of bladder volume. The standard method of ruling out post-obstructive uropathy is straight catheterization. Hemodynamically stable patients may go up to 6 h without urination before undergoing straight catheterization to rule out obstruction.

In patients with a small volume of urine, further workup of postrenal causes between the kidney and the bladder should be evaluated. Prerenal causes should also be ruled out. Ultrasound of the retroperitoneum should be urgently obtained to rule out hydronephrosis. Therapy may include ureteral stenting or percutaneous nephrostomy tube placement in addition to removal of the blockage.

Initial treatment for oliguria should focus on reversing the underlying cause and correcting any fluid or electrolyte imbalances before acute renal failure occurs. Focus should be placed on postrenal and prerenal etiologies first. Volume status should be ascertained quickly. Hypovolemic patients should be given isotonic intravenous fluids, while hypervolemic patients should undergo diuresis.

Hypervolemia presents with jugular venous distention, an S3 gallop, pulmonary rales, and/or peripheral edema. Invasive monitoring of the central venous pressure and the cardiac output are useful adjuncts in monitoring the fluid status of a patient. While there is no evidence to support low-dose dopamine for recovery of acute renal failure, with adequate preload, it is important to augment the blood pressure if needed to maintain a MAP of 60 mmHg or higher.

In patients where initial therapy does not halt the progression of renal failure, renal consultation is recommended. Indications for dialysis include hyperkalemia, acidosis, hypervolemia, and uremia.

85
Postoperative Fever

Kristin Gross

Introduction

Postoperative fever is defined as a temperature greater than 101.3 °F. In its most basic form, fever is the manifestation of specific cytokine release, most commonly IL-1, IL-6, TNF-alpha, and IFN-gamma. These cytokines are released by tissue trauma and *do not necessarily signal infection*. Most early postoperative fevers (postoperative days 1–2) are caused by the inflammatory stimulus of surgery and resolve spontaneously. However, efflux of these cytokines is also stimulated by bacterial endotoxins and exotoxins possibly indicative of an infectious cause.

The timing of fever relative to the day of surgery provides a consistent and formulaic approach to generating a differential diagnosis. The classic pneumonic known as the "5 Ws" of postoperative fever provides the most likely cause of a fever based on postoperative day (POD) as follows:

- *Wind* – atelectasis on POD 1–2
- *Water* – urinary tract infection on POD 3–5
- *Walking* – deep venous thrombosis (DVT) or pulmonary embolism (PE) on POD 4–6
- *Wound* – wound infection on POD 5–7
- *Wonder drugs* – drug-related fever on POD >7

Although the "5 Ws" provide an excellent and memorable approach to the differential diagnosis of postoperative fever, the following algorithm based on timing incorporates the most likely etiologies of postoperative fevers based on four time periods: immediate, acute, subacute, and delayed (Fig. 85.1).

Immediate

This is defined as fever intraoperatively or within the first 24 h after surgery. Fever during the intraoperative period may be due to endocrine disturbances (Addisonian crisis, thyroid storm, or pheochromocytoma), transfusion reaction, drug hypersensitivity, preexisting infection, intraoperative manipulation of infected or necrotic material, or malignant hyperthermia. Fevers during the first 24 h after surgery are most often due to the normal inflammatory response to surgery. However, an important and critical etiology to recognize during this time frame is necrotizing wound infection secondary to *Streptococcus pyogenes* (group A) or *Clostridium perfringens*. Patients with these infections typically spike high fevers (103–104 °F) and have painful wounds. A bedside evaluation of the wound is essential in this setting, inspecting for ominous changes such as discoloration, intense erythema, crepitus, and foul-smelling brown drainage. Prompt recognition is crucial, as treatment requires emergent operative debridement and appropriate antibiotics.

Acute

This is defined as fever within the first week of surgery (POD 1–7). Fever during this time frame is overall *most commonly due to infections*. Although atelectasis is commonly noted as a common cause of fever on postoperative days 1–2, some argue that the concurrence is more likely coincidental and not causal and ultimately related to the inflammatory and stress state that defines most postoperative periods. Fevers on postoperative days 3–5 are commonly due to urinary tract infections, particularly in patients with indwelling urinary catheters. The diagnosis can be made quickly by urinalysis. Based on cultures, a urinary tract infection is defined as a urine culture growing >100,000 colonies of bacteria. A persistent fever and respiratory symptoms such as tachypnea, shortness of breath, and infiltrates on chest X-ray should raise suspicion for pneumonia. Particular populations at risk include those on the ventilator (ventilator-associated pneumonia, VAP) and those with decreased gag reflex (aspiration pneumonia). Sputum cultures may be obtained to elucidate a specific pathogen. Fever on postoperative days 5–7 is likely to represent surgical site infection; the most common location is the wound itself. Tenderness, induration, erythema, and drainage from the incision site are all indicators of a

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_85, © Springer New York 2015

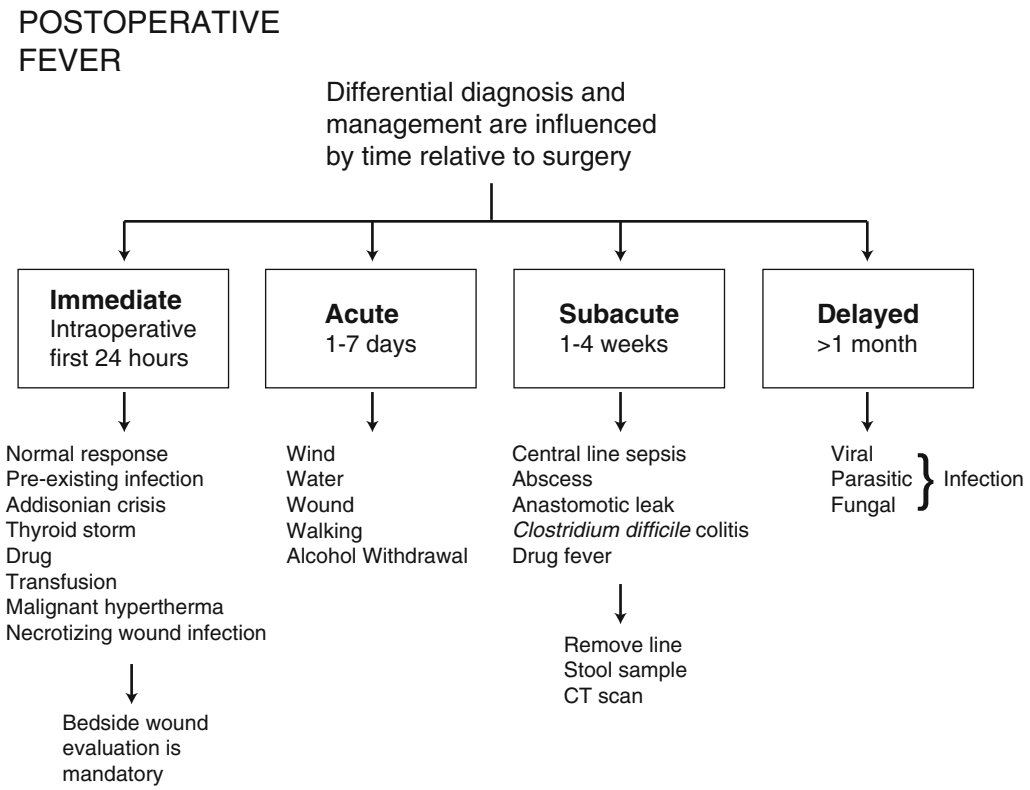

FIG. 85.1 Treatment algorithm for postoperative fever. *CT* computed tomography

possible wound infection. Specifically in a patient who underwent abdominal surgery, intra-abdominal abscess or anastomotic leak should also be strongly suspected. These diagnoses may be investigated with computed tomography (CT) scan of the abdomen/pelvis with oral and intravenous contrast.

Noninfectious causes of fever within the first week of surgery include deep venous thrombosis/pulmonary embolism (typically postoperative days 4–6), alcohol withdrawal (typically postoperative day 3), and myocardial infarction. In the bed-bound patient with no obvious infectious sources of fever, thromboembolism should be considered and assessed with lower extremity Doppler ultrasound and potentially a spiral chest CT with intravenous contrast. In the setting of hemodynamic instability, mental status changes, and hallucinations, alcohol withdrawal is a potential cause of fever.

Subacute

This is defined as fever 1–4 weeks following surgery. The most common infectious causes of postoperative fever during the subacute period are blood catheter-related infections and surgical site infections. The risk of central line sepsis increases with the duration of the indwelling catheter. *Thus, a persistent fever and/or leukocytosis >1 week following surgery in a patient with a central line is most likely related to the catheter*. As patients get further away from their initial surgical encounter, the risk of antibiotic-associated diarrhea caused by *Clostridium difficile* increases while the duration of antibiotic therapy increases. Patients with fever and diarrhea should undergo stool cultures and *C. difficile* toxin assay. Postoperative day seven is a particularly common time to present with an anastomotic leak in the abdominal surgery patient. Other clues to anastomotic leak besides fever include tachycardia, pain, diarrhea, persistent ileus, and leukocytosis. Common noninfectious causes of fever after the first week of surgery are thromboembolic events and febrile drug reactions. Antibiotics are the most common cause of drug fevers.

Delayed

This is defined as fever greater than 1 month after surgery. At this point, the risks of noninfectious causes of postoperative fever previously described are all likely decreased. In the delayed period, patients may develop more rare infections due to viral, parasitic, and fungal pathogens.

86
Postoperative Chest Pain

Elizabeth Berger

Introduction

Chest pain can originate from various anatomical structures in the body. The way in which chest pain presents in terms of location, duration, and character can help distinguish the source of the pain: cardiac, pulmonary, gastrointestinal, vascular, or musculoskeletal (Fig. 86.1). The workup for chest pain in the postoperative period should typically mirror that of an emergency room workup with the additional consideration of incisional pain.

Patient History

In order to assess postoperative chest pain, one should know the patient's preoperative history, the procedure that was performed, and when it occurred. In reviewing a patient's preoperative history, certain cardiovascular risk factors can be identified, such as hypertension, hypercholesterolemia, or diabetes. There are pulmonary and gastrointestinal risk factors that can contribute to postoperative chest pain, such as chronic obstructive pulmonary disease (COPD), gastroesophageal reflux disease (GERD), or peptic ulcer disease (PUD). A history of cholelithiasis could also present as postoperative chest pain in the setting of narcotic use. Typically, chest pain in the *immediate* postoperative period is most often cardiac related, whereas infectious, thromboembolic, and pulmonary causes of chest pain occur later in the postoperative setting.

Initial Evaluation of Patient

If a clinician is called to evaluate postoperative chest pain, a complete history and a new set of vitals should be taken immediately. During this initial assessment, a patient should be placed on the appropriate monitors and overall stability should be determined. A pulse oximeter, telemetry, and a blood pressure cuff should be placed. The patient might be better served in a more monitored setting such as an intensive care unit. As the patient's history is being taken, a complete physical exam should be completed simultaneously. The exam should listen for new murmurs, friction rubs or gallops, presence or absence of breath sounds, and abdominal tenderness. The surgical wound and other drains should be assessed for change in character or consistency. Initial tests that are quick to do at the bedside should be completed. These include an electrocardiogram (EKG) and cardiac enzymes to evaluate a possible cardiac cause, an arterial blood gas and chest X-ray to assess pulmonary status, and a CBC and complete metabolic profile for any possible intra-abdominal problems.

Cardiac Etiology of Chest Pain

A cardiac cause of chest pain could quickly become fatal if not adequately addressed in a timely manner. Symptomatic ischemic heart disease can present as either angina pectoris or a myocardial infarction (MI). The recently postoperative patient who has sudden onset chest pressure that is constant, sometimes associated with diaphoresis, may be experiencing either angina or MI. In this circumstance, an EKG should be done and cardiac enzymes should be drawn immediately. Oxygen should be placed on the patient and morphine can be given to relieve the pain. Chest pain occurring in the setting of EKG changes, such as elevated ET segments and inverted T waves and Q waves, or positive troponin levels justifies a diagnosis of myocardial infarction. Three troponin levels obtained 6 h apart are highly sensitive and specific for detecting myocardial injury. To treat the actual chest pain if it is suspected to be cardiac in nature, *sublingual nitroglycerin* (0.4 mg every 5 min) can relieve ischemic pain of angina but not the pain of an MI. *Aspirin should also be given in the acute setting. Beta antagonists* can be given as well, unless contraindicated (heart failure, bradycardia, heart block, or COPD). In assessing a patient with postoperative chest pain that is likely cardiac in nature, one should

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_86, © Springer New York 2015

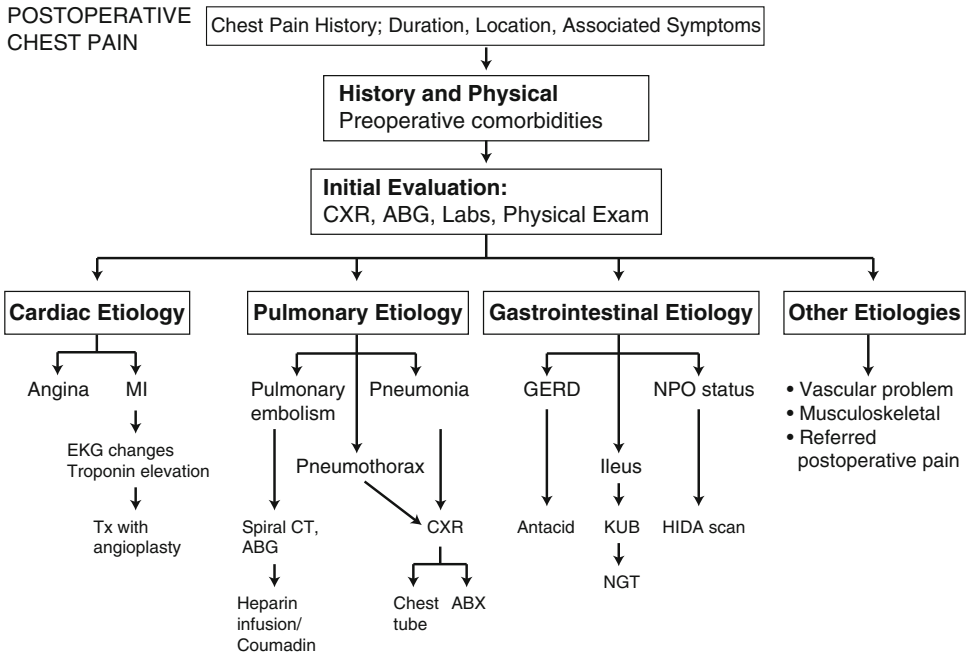

FIG. 86.1 Algorithm for postsurgical chest pain. *CXR* chest X-ray, *ABG* arterial blood gas, *MI* myocardial infarction, *GERD* gastroesophageal reflux disease, *NPO* nil per os, *CT* computed tomography, *ABX* antibiotics, *HIDA* hepatobiliary, *KUB* kidneys, ureters, and bladder, *NGT* nasogastric tube

keep in mind that an MI occurs much less frequently after postoperative day 3, chest pain is present in only 30 % of postoperative myocardial infarctions, and thrombolytic therapy is not indicated if an early MI is encountered (<6 h). *Angioplasty is the treatment of choice for the patient who experiences a postoperative MI.*

Pulmonary

Cardiac pain is centrally located while inflammation of the parietal or diaphragmatic pleuras may present more laterally. Pulmonary causes of chest pain tend to be associated with coughing, breathing, or shortness of breath. The etiologies to consider in the postoperative period are pulmonary embolus, pneumothorax, pneumonia, or pleurisy. A pneumothorax tends to occur in the immediate postoperative period, especially in upper abdominal surgery or laparoscopy. The presentation of a large pneumothorax is often sudden onset of shortness of breath associated with sharp, pleuritic chest pain. An arterial blood gas and chest X-ray should be done to rule out pulmonary etiology in a postoperative patient with chest pain. Pneumothoraces are identifiable on chest X-ray.

A pulmonary embolus (PE) may occur several days into the postsurgical period. Risk factors for PE include prior history of deep venous thrombosis (DVT), obesity, prolonged periods of immobilization, cancer, and long operative times. If a clinician evaluates a patient with acute onset of shortness of breath, chest pain, tachycardia, and diaphoresis, pulmonary embolus should be high on the differential list. An arterial blood gas typically reveals hypoxemia associated with a respiratory acidosis (hypercarbia). If the pulmonary embolus is large enough to cause right heart strain, one could see EKG changes such as right axis deviation. Further diagnostic studies include a *spiral CT scan of the chest, which is the standard diagnostic test*. If a patient has poor kidney function, a ventilation perfusion scan can be performed in lieu of a CT scan. If a clot is found on CT scan, treatment involves anticoagulation (typically a heparin drip bridged to Coumadin) if the patient is greater than 24 h postsurgery. Placement of an inferior vena cava filter should also be considered in postoperative patients as well as pulmonary embolectomy in rare cases.

Pneumonia can also cause chest pain in the postoperative setting. Like a pulmonary embolus, pneumonia often occurs several days after surgery and can occur in up to 20 % of patients. Typically, atelectasis progresses to pneumonia, which can often present with chest pain, fevers, labored breathing, and a leukocytosis. Antibiotics are typically the treatment of choice for pneumonia.

Gastrointestinal Causes

Gastrointestinal causes of chest pain should not be excluded when assessing a postsurgical patient. The stomach and duodenum can be potential sources of referred pain to the chest if there is inflammation or ulceration present. If a patient

complains of epigastric pain that is exacerbated by laying flat, adequate antacid therapy should be initiated in attempts to relieve possible gastroesophageal reflux symptoms. If a patient has been nil per os (NPO) for an extended period of time, biliary stasis can occur, which can lead to acalculous cholecystitis. Likewise, if a patient begins to eat and has multiple episodes of emesis, Boerhaave's syndrome should be considered and a chest X-ray should be ordered to assess for possible mediastinitis. A prolonged ileus can also cause abdominal distension, which can cause chest discomfort from a distended stomach.

Less Common Causes

Complications from surgery as well as certain vascular disorders can cause postoperative chest pain that are less common than the various etiologies outlined previously. If a patient has bad cardiovascular disease and develops sudden, sharp chest pain that radiates to the back, one should consider a ruptured or dissecting aortic aneurysm. Vital signs should be monitored extremely carefully if this is suspected. Referred pain after laparoscopy, an anastomotic leak in the abdomen, or diaphragmatic abscess can also cause chest discomfort. There is always the possibility that postoperative chest pain is musculoskeletal in nature.

Conclusion

Postoperative chest pain is a significant symptom in patients and needs to be evaluated with promptness and thoroughness. One should have an algorithm in mind when assessing a postsurgical patient who complains of chest pain. Fatal complications should always be a top priority and should be ruled out before other, less-threatening etiologies are considered. A postsurgical patient with chest pain should never be diagnosed with anxiety or musculoskeletal pain until other causes are ruled out.

87
Ventilator Management

Vikram D. Krishnamurthy

Introduction

Respiratory failure is defined as inadequate gas exchange by the respiratory system and can be a result of inadequate *oxygenation* or *ventilation. Oxygenation* is defined as the delivery of oxygen to tissues for consumption after inhaled oxygen has diffused into the blood oxygen content. This relies on the hemoglobin concentration, oxygen saturation, and dissolved portion of oxygen in the blood. Oxygenation is commonly quantified using the PaO_2 from blood gas measurements or the oxygen (O_2) saturation recorded from pulse oximetry. *Ventilation* measures the adequacy of carbon dioxide (CO_2) elimination. CO_2 is produced when tissues utilize fuel in the presence of oxygen and the amount of CO_2 produced is dependent on the respiratory quotient of the substrate.

Respiratory Failure

As previously mentioned, respiratory failure results from inadequate oxygenation or ventilation. Inadequate oxygenation may be due to the lack of systemic oxygen delivery, a ventilation-perfusion (V/Q) mismatch (shunting), or hypoventilation. It is defined as a PaO_2 less than 60 mmHg or O_2 saturation of less than 90 %. Alternatively, insufficient ventilation is related to a reduced ventilatory drive resulting from neurologic disorders, muscular weakness, or sedating medications. Ventilation is monitored using the patient's respiratory rate (RR), tidal volume (V_T), blood gas pH, and PCO_2.

Indications for Mechanical Ventilation

Criteria for intubation rely on clinical and biochemical factors. Decline in mental status, inability to protect the airway, and increased work of breathing are common clinical factors that escalate the need for mechanical ventilation (Fig. 87.1). Biochemical factors include a persistent PaO_2 value of <70 despite the delivery of 100 % oxygen or CO_2 retention as evident by a sustained elevated PCO_2. The classic adage is to intubate as soon as the clinician has begun to entertain the notion, as it may prevent delay in supportive care, diagnosis, and potential treatment.

Ventilator Settings and Management

Providing mechanical ventilation allows the clinician control over several settings including RR, V_T, positive end-expiratory pressure (PEEP), and the fraction of inspired oxygen (FIO_2). Oxygenation is affected by the PEEP and FIO_2. Terminal airways can collapse with expiration and the application of PEEP prevents this by stenting these airways open. The FIO_2 of room air is 21 % but the ventilator can deliver FIO_2 in increasing amounts up to 100 %. Ventilation can be affected by manipulating the RR and V_T as minute ventilation (MV) is derived from these two parameters ($MV = RR \times V_T$). In the majority of clinical scenarios, the initial ventilator settings are a RR of 16–18 breaths/min, V_T of 7–12 mL/kg (using ideal body weight), PEEP of +5, and FIO_2 of 100 %. It is important to note that different settings may be employed and parameters set when treating with specific diseases such as acute respiratory distress syndrome (ARDS), which will not be discussed here.

Changes in the ventilator settings are largely driven by patient factors and blood gas measurements. Ventilator settings are titrated to minimize *volutrauma*, *barotrauma*, and O_2 toxicity while keeping the patient comfortable utilizing sedation strategies appropriate to the patient's expected clinical course. Volutrauma results from inflation-related damage to the alveoli as opposed to barotrauma, which is caused from airway pressure damaging the alveoli. These two effects can be mitigated by appropriately altering the ventilator mode, RR, $V_{T,}$ or PEEP. Oxygen toxicity refers to the observation that oxygen can be deleterious when delivered at high concentrations for extended periods of time and this can be avoided by limiting unnecessary FIO_2 and PEEP.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_87, © Springer New York 2015

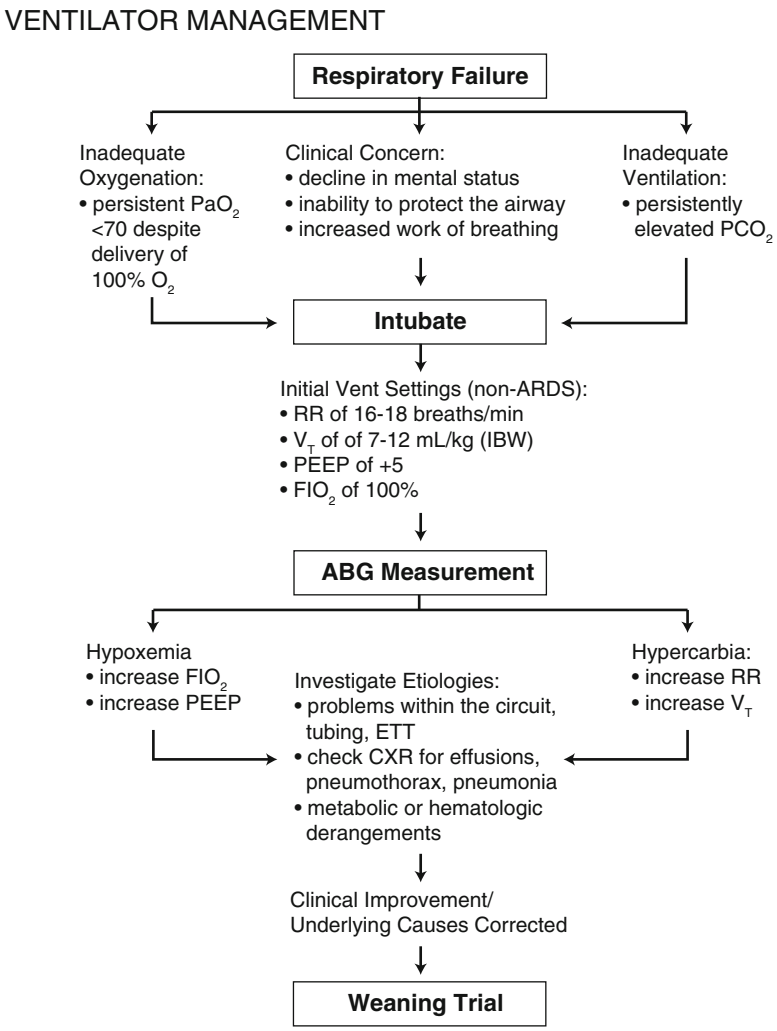

FIG. 87.1 Respiratory failure and ventilator management. *ARDS* acute respiratory distress syndrome, *RR* respiratory rate, V_T tidal volume, *IBW* ideal body weight, *PEEP* positive end-expiratory pressure, FIO_2 fraction of inspired oxygen, *ETT* endotracheal tube, *CXR* chest X-ray

In summary, once the patient is mechanically intubated, the following basic algorithm may be used: Problems with oxygenation as evident by an undesired PaO_2 or O_2 saturation can be addressed by altering the FIO_2 or PEEP. Problems with ventilation as evident by an undesired pH or PCO_2 can be addressed by altering the RR or V_T.

In the mechanically ventilated patient, derangements in either oxygenation or ventilation must be investigated only after obvious malfunctions with the extracorporeal systems have been excluded. For example, the placement and patency of the endotracheal tube, the ventilator tubing, and the pulse oximeter must all be evaluated when clarifying the causation of perceived changes in respiratory mechanics.

Discontinuation of Therapy

Mechanical ventilation in most situations should be considered as temporary and supportive until the underlying clinical factors that necessitated intubation are being treated and corrected. The concept of "weaning" from the ventilator involves limiting the patient's dependence on the services it provides. For example, a patient must be awake and able to breathe spontaneously before entertaining extubation. Furthermore, oxygen requirements must be trivial as evident by a minimal amount of FIO_2 or PEEP required to maintain an adequate PaO_2 or O_2 saturation. Once these basic criteria are met, then the patient undergoes a spontaneous breathing trial (SBT).

The ventilator is set so that the PEEP is +5, the FIO_2 is <40 %, and neither RR nor V_T is provided. Continuous positive airway pressure (CPAP) is provided and the patient's clinical response, O_2 saturation, RR, and V_T are monitored. If the patient remains comfortable, can maintain adequate oxygenation, does not become tachypneic nor apneic, and can generate adequate V_T, then he/she is a candidate for extubation.

The rapid-shallow breathing index (RSBI) is frequently used while performing an SBT. The RSBI is calculated by dividing the RR by the V_T, provided as a fraction of 1,000 mL (i.e., a V_T of 500 mL would be entered into the denominator

as 0.5). Intuitively, the lower the RSBI, the more successful the patient is at breathing without the ventilator, as this would require that their RR is low and the V_T is high. Thus, an RSBI of less than 105 is frequently used as a component to determine successful extubation.

Failure to wean from the ventilator is a complex topic that is affected by many variables and can be treated by differing approaches. Reasons for failed extubation include, but are not limited to, factors related to organic neurologic disorders, sedation, chest wall compliance, muscular fatigue, pleural effusions, and pneumonia. Identifying and treating the potential underlying causes is imperative. In some cases, progressive weaning can be carried out with T-piece trials or slowly de-escalating pressure support. Ultimately, the prevailing goal is to limit ventilator support and to extubate as early as possible, thus avoiding the adverse outcomes of endotracheal intubation (i.e., pneumonia, patient discomfort, suboptimal oral hygiene, vocal cord damage, laryngomalacia).

88
Postoperative Hypoxia and Pulmonary Embolism

Patri Marconi

Introduction

In the postoperative period, adequate oxygen delivery to the tissues is necessary to ensure proper healing and recovery. Failure to do so results in tissue *hypoxia*. This can be due to poor oxygen delivery to the tissues or poor utilization of oxygen by the tissues. When tissues become hypoxic, aerobic metabolism converts to anaerobic metabolism. This ultimately provides an unsustainable environment for cells, causing cell death. It is imperative that physicians are able to understand the causes of tissue hypoxia, the presenting symptoms, and how to treat these causes to avoid life-threatening consequences (Fig. 88.1). The development of deep vein thrombosis (DVT) resulting in pulmonary embolism (PE) as a cause of hypoxia is of particular concern in postoperative patients and will be discussed in greater detail.

Main Causes of Hypoxia

Hypoxemia

Hypoxemia develops when the oxygen tension in the arterial blood (P_{aO2}) is low, signifying an inability of the lungs to adequately oxygenate the blood. This can be a result of:

1. Hypoventilation causing an increased P_{CO2} (excessive narcotic use, residual anesthesia effects)
2. V/Q (ventilation–perfusion) mismatch from decreased alveolar filling (e.g., pneumonia, atelectasis) or increased pulmonary capillary pressure (e.g., pulmonary edema from fluid overload or heart failure)
3. Pulmonary shunting causing perfusion without gas exchange (e.g., acute respiratory distress syndrome (ARDS), fat embolization, smoke or chemical inhalation)
4. Pulmonary embolism causing infarction of the peripheral lung tissue

Arterial P_{O2} is measured with an arterial blood gas (ABG) and used in conjunction with the alveolar oxygen tension (P_{aO2}) to determine the A-a (alveolar–arterial) gradient. P_{AO2} is calculated using the following equation:

$$P_{AO2} = F_{IO2}\left(P_{atm} - P_{H2O}\right) - \left(P_{aCO2} / R\right)$$

where F_{IO2} is the fraction of inspired oxygen, P_{atm} is the atmospheric pressure at sea level (760 mmHg), P_{H2O} is the pressure of water vapor in inspired air (47 mmHg), P_{CO2} is the carbon dioxide tension in arterial blood, and R is the respiratory quotient (estimated at 0.8). The A-a gradient is calculated using the following equation:

$$P_{aCO2} \div P_{AO2}$$

A normal A-a gradient is 0.77–0.82. A widened A-a gradient would imply a V/Q mismatch that should improve with increased administration of oxygen (increasing the oxygen tension in the alveoli will decrease the gradient and improve oxygenation of blood).

Anemia

Decreased hemoglobin levels cause a decrease in the oxygen-carrying capacity of blood and, therefore, decrease tissue oxygenation.

Decreased Circulation

Impaired cardiac output (i.e., cardiogenic shock) does not allow proper delivery of oxygen to the tissues. Shunting of

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_88, © Springer New York 2015

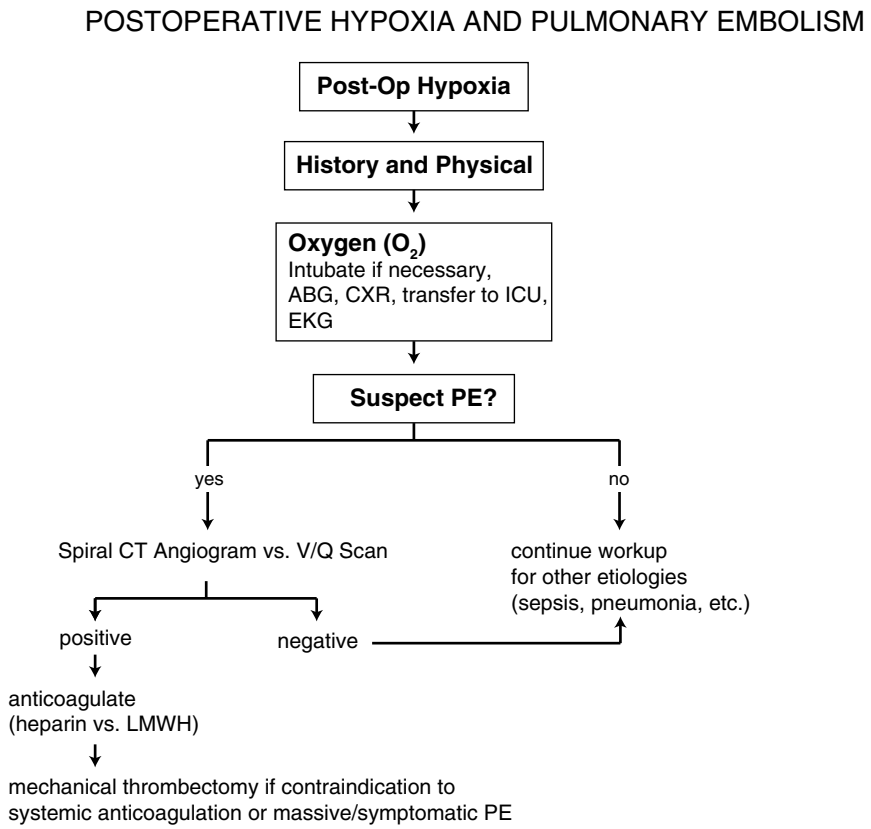

FIG. 88.1 Treatment algorithm for postoperative hypoxia and pulmonary embolism (*PE*). *ABG* arterial blood gas, *CXR* chest X-ray, *EKG* electrocardiogram, *LMWH* low molecular weight heparin

blood that causes the lungs to be bypassed (i.e., cirrhosis) will result in decreased oxygenation of the blood.

Increased Oxygen Affinity

Shift of the oxygen dissociation curve to the left will result in increased affinity of oxygen to hemoglobin molecules and subsequent decreased delivery to the tissues.

Pulmonary Emboli

Pulmonary emboli (PEs) are most commonly the result of thrombi in the deep veins of the lower extremities (deep vein thromboses or DVTs). Large PEs can cause right heart strain and/or sudden death by blocking blood flow through the main pulmonary arteries. Smaller PEs can travel peripherally and may cause infarctions in the lungs. All of these will cause a V/Q mismatch and can cause minor to severe hypoxia.

Presentation

When presented with a patient with decreasing oxygenation, a proper history is essential to help determine the cause. Patients in the immediate postoperative period are more likely to have hypoxia as a result of atelectasis or anesthesia/analgesic effects. Poorly controlled pain, excessive pain medication use, and level of alertness should all be evaluated. Pneumonia as a cause of hypoxia is more likely later in the patient's postoperative course. A history of restrictive or obstructive pulmonary disease (asthma, chronic obstructive pulmonary disease (COPD)), smoking history, cardiac disease history, or a recent history of respiratory illness should all be ascertained as well. A family history of clotting disorders or a personal history of DVTs or miscarriages should raise suspicion for a PE being the cause of hypoxia as a result of an undiagnosed hypercoagulable state.

The most common first signs and symptoms of hypoxia are a decrease in oxygen saturation, tachypnea, tachycardia, and dyspnea. If a patient has cardiac dysfunction at baseline, tachycardia may not be the typical presentation. As the degree of hypoxia and hypoxemia worsen, other signs such as a decline in mental status, cyanosis of the extremities, and bradycardia may develop. Early identification and treatment of hypoxia is essential to avoid lasting damage to the patient.

DVTs may present with lower extremity swelling, pain, and/or erythema, but may have no presenting symptoms at all. Homan's sign (pain with passive dorsiflexion of the ankle) has been noted in patients with DVTs, but is neither sensitive nor specific. PEs present with nonspecific signs that may be confounded by prior cardiac or pulmonary

diseases. Such signs and symptoms include tachypnea, tachycardia, crackles on auscultation, dyspnea, and pleuritic chest pain.

Assessment/Treatment

The initial step in the assessment of a hypoxic patient is to determine the severity of their condition. In an unstable patient, immediate delivery of oxygen is necessary. The physician needs to determine if intubation is indicated based on the patient's status and response to high-flow oxygen (nonrebreather or other noninvasive delivery methods). Simultaneously, an ABG and blood lactate should be drawn and a chest X-ray done. The patient should be transferred to an intensive care unit for closer monitoring and further workup. Sepsis should be ruled out with cultures of all appropriate lines and wounds and of the urine. Broad-spectrum antibiotics should be started after cultures are drawn if the suspicion for sepsis is high. An electrocardiogram (EKG) and echocardiogram should be done to look for a cardiac cause of hypoxemia or to look for signs of right heart strain that may be indicative of a PE.

If clinical suspicion for PE is high, even without findings consistent with right heart strain on EKG or echocardiogram, spiral CT angiography or (less ideally) a ventilation and perfusion (V/Q) scan should be done. While a normal result on a V/Q scan will exclude PE and a high-probability result is specific for a PE, the majority of results will have an intermediate result, making V/Q scanning less helpful. The drawbacks of CT angiography are the need for an intravenous contrast dye load (which is contraindicated in patients with renal failure) and its low sensitivity for small peripheral emboli. However, it has a high negative predictive value and high specificity.

Initial treatment for a PE is anticoagulation. Heparin therapy should be started and the decision to give unfractionated versus low molecular weight heparin (LMWH) should be made by the physician. Unfractionated heparin requires a continuous intravenous infusion with serial checks of the aPTT (activated partial thromboplastin time) to avoid excessive anticoagulation, but its effect in the blood is reversed fairly rapidly when the infusion is stopped. LMWH is available in daily or twice-daily subcutaneous dosing and does not require monitoring given its weight-based dosing, but its effects remain for longer than that of unfractionated heparin. Long-term therapy with warfarin should begin while the patient is on heparin to avoid the initial hypercoagulable state seen with warfarin dosing alone. Anticoagulation treatment for a postoperative DVT or PE should last for 3 months. Treatment for a hypercoagulable disorder or idiopathic DVT or PE should be 6–12 months. Lifelong anticoagulation should be considered if a patient has had multiple DVTs/ PEs.

Mechanical thrombectomy has been performed in patients with contraindications to systemic anticoagulation or thrombolysis and have a massive or symptomatic PE. The method used (percutaneous versus surgical) is determined by the patient's stability and the immediate availability of an interventional radiologist or a cardiac surgeon.

In a stable patient, the oxygen saturation and serum bicarbonate levels should be checked. If the oxygen saturation is greater than 95 % and bicarbonate levels are normal, the patient can likely be monitored without further testing. If the oxygen saturation is low but greater than 90 %, low-flow oxygen through a nasal cannula should be administered and a chest X-ray performed. If the oxygen saturation does not improve (or drops below 90 %) or if the bicarbonate level is excessively high or low (indicative of an acid–base abnormality), the physician should proceed with a chest X-ray, ABG, and lactate level. Given these results, further testing may be necessary.

89
Postoperative Hypotension

Shaun Daly

Evaluation

The initial evaluation of a patient with postoperative hypotension involves the *immediate exclusion of life-threatening treatable causes and initiation of early resuscitation* (Fig. 89.1). One must verify the accuracy of the blood pressure reading before early and aggressive measures are instituted. A cuff larger than two-thirds the circumference of the arm or an improperly calibrated arterial line may give falsely low blood pressure readings. Once the determination has been made that the reading is accurate, supplemental oxygen and fluid resuscitation with crystalloid need to be urgently initiated. Airway, breathing, and circulation (ABC) should be promptly assessed in the unstable patient. One should place the patient in Trendelenburg position. A rapid, accurate patient history needs to be obtained with particular attention paid to home and current medications, allergies, perioperative and intraoperative events (e.g., blood loss), the circumstances surrounding the current hypotensive episode (rapidity of onset, associated symptoms, etc.), and any use of intraoperative stress dose steroids. Physical exam should include vital signs (blood pressure, pulse, respiratory rate, temperature), oxygen saturation, and hourly urine output. The patient's mental status is determined and compared to known baseline. Secondary survey should include a thorough heart, lung, and pulse exam. The surgical wounds and drains need to be evaluated.

Workup

Proper laboratory assessment is critical and should include an arterial blood gas, complete blood count, basic metabolic profile, coagulation profile, and a type and screen. An electrocardiogram, troponin level, and chest X-ray need to be obtained. Invasive monitoring needs to be considered in an unstable patient and in a patient who does not respond appropriately to resuscitation. A Foley catheter needs to be placed. Additional invasive monitoring techniques available include arterial line blood pressure monitoring, a central venous catheter, and a Swan-Ganz catheter to provide objective data regarding heart function.

Differential Diagnosis

Life-Threatening Hypotension

Life-threatening treatable causes of hypotension need to be excluded immediately. A **tension pneumothorax** restricts venous blood flow to the heart and causes acute hypotension. Immediate needle decompression with a large-gauge needle in the second intercostal space midclavicular line is indicated without delay. After needle decompression, a chest tube is required for definitive treatment. **Cardiac tamponade** results in equalization of diastolic pressures of the heart and acute hypotension. Only low volumes of blood are needed to cause life-threatening hypotension in acute tamponade. An immediate bedside pericardiocentesis is indicated, followed by a formal pericardial window in the operating room for definitive treatment. A **pulmonary embolism** needs to be excluded.

Decreased Preload

Decreased systemic blood pressure can be the result of decreased preload. Hemorrhage, *the most common reason for postoperative hypotension*, must be recognized promptly and acted on in an expedited manner. External hemorrhage can be controlled with bedside pressure or ligation of the bleeding source. Internal bleeding causing hypotension usually requires reoperation to control; however, it is reasonable to first check the patient's coagulation profile. A patient with an abnormal coagulation profile needs to be treated with blood products to correct the coagulopathy,

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_89, © Springer New York 2015

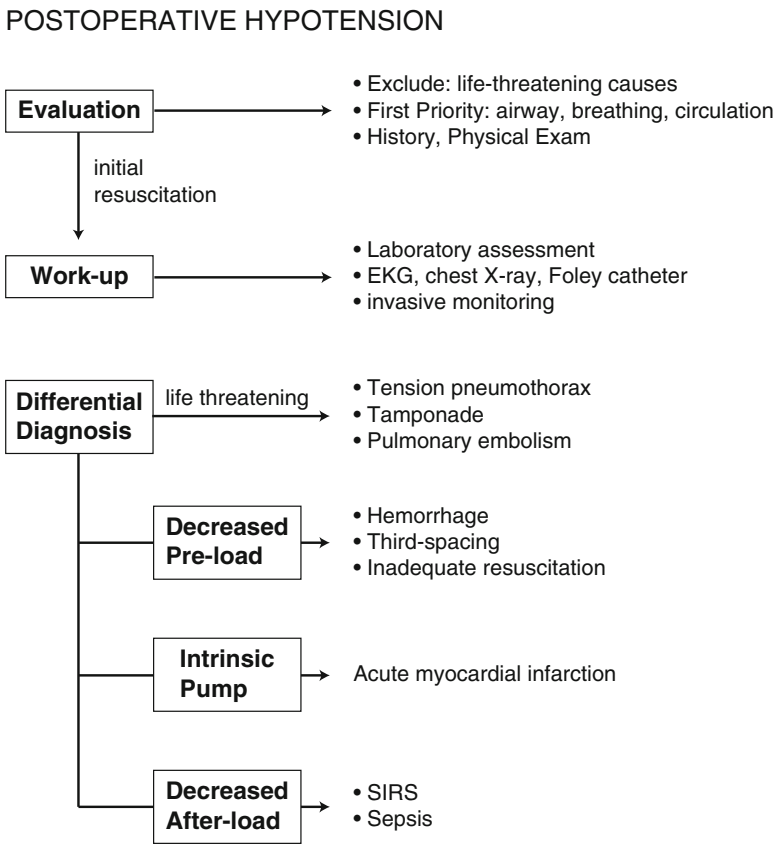

FIG. 89.1 Treatment algorithm for postoperative hypotension. *EKG* electrocardiogram, *SIRS* systemic inflammatory response syndrome

reserving a return to the operating room for a failure of the blood products to control bleeding. Even in the presence of an abnormal profile, the threshold for returning to the operating room should be low. Two additional common causes for decreased preload include third spacing of fluids and inadequate intraoperative fluid replacement. Fluid resuscitation is the mainstay of therapy for these causes of postoperative hypotension. Additional diagnoses include sepsis, burns, high epidurals, and drug-related and high-PEEP mechanical ventilation.

Intrinsic Pump Malfunction

Decreased systemic blood pressure can be the result of intrinsic pump failure, a common cause of which is an acute myocardial infarction. Patients with postoperative hypotension should be evaluated with an electrocardiogram and troponin levels. Additional causes of intrinsic pump malfunction include dysrhythmias, cardiomyopathies, valvular disorders, cardiac depressant drugs, and decompensated congestive heart failure. A patient with intrinsic pump failure needs invasive monitoring and treatment needs to be in response to the objective data invasive monitoring provides.

Decreased Afterload

Decreased blood pressure can be the result of decreased afterload. A common cause of decreased afterload is the vasodilatory effects of the systemic inflammatory response syndrome (SIRS) to surgery and sepsis, if infection is present (see Chap. 91). Sepsis requires prompt treatment with broad-spectrum antibiotics and aggressive fluid resuscitation guided by invasive monitoring. Strict algorithmic guidelines for the treatment of sepsis can be found via the Surviving Sepsis Campaign.[1] Sources of immediate sepsis are necrotizing infections of the fascia and soft tissue, mandating evaluation of all surgical wounds in a postoperative patient with hypotension. Anaphylaxis is a less common cause for postoperative hypotension but must be recognized because emergent treatment with epinephrine is essential. Additional causes of decreased afterload-induced postoperative hypotension are neurogenic shock, high epidurals, burns, and drug-related and transfusion-related reactions.

[1] http://www.survivingsepsis.org

90
Necrotizing Soft Tissue Infections

Andrew T. Arndt

Introduction

Necrotizing soft tissue infections (NSTIs) are severe bacterial infections of the soft tissues, characterized by necrosis of the skin, subcutaneous tissue, fascia, or muscle. They are rapidly progressive and often fatal; as such, prompt recognition and early initiation of therapy – namely, aggressive surgical debridement – are crucial steps for improving survival (Fig. 90.1). NSTIs progress from deep infections to necrosis by microvascular thromboses caused by the bacteria and the infiltration of polymorphonuclear cells. The tenuous blood supply of the subcutaneous tissue and fascia predisposes to the spread of infection. Additionally, some organisms like *Clostridium perfringens* produce toxins that cause necrosis of well-perfused tissues.

Etiology

Although the mainstay of therapy for NSTIs is surgical debridement, antibiotic therapy also plays an important role in treatment. Therefore, understanding the pathogens responsible for NSTIs is of key importance. NSTIs are classified as either **polymicrobial** or **monomicrobial**. Most NSTIs are polymicrobial, with an average of 4.4 organisms isolated. *Monomicrobial infections tend to be more severe and aggressive and consequently carry a greater mortality.*

Polymicrobial Infections

Polymicrobial NSTIs involve mixed aerobes and anaerobes and are typically found in the perineal and perirectal areas, in chronic diabetic and pressure ulcers of the lower extremities, and at surgical sites. They typically consist of both skin and fecal flora, including *Staphylococcus* species, *Streptococcus* species, *Enterococcus* species, gram-negative enteric bacilli, and anaerobes. NSTIs arising from bite wounds are typically polymicrobial and involve unique organisms, such as *Pasteurella* species and *Capnocytophaga canimorsus* in animal bites and *Haemophilus* species and *Eikenella corrodens* in human bite wounds.

Monomicrobial Infections

Monomicrobial NSTIs tend to be rapidly progressive and carry a high mortality rate. The most typical pathogens in monomicrobial NSTIs are *Streptococcus pyogenes* (group A β-hemolytic *Streptococcus*), *Clostridium* species, and, particularly in recent years, methicillin-resistant *Staphylococcus aureus* (MRSA). Group A β-hemolytic *Streptococcus* (GAS) can produce virulence factors and exotoxins that lead to the rapid spread of infection and necrosis in nonischemic tissue. *Clostridium perfringens* accounts for roughly 80 % of clostridial NSTIs and can lead to a rapidly progressive clostridial myonecrosis (gas gangrene), in which potent extracellular toxins lead to direct tissue injury as well as hemolysis and microvascular thrombosis. Clostridial myonecrosis classically occurs with traumatic puncture; in recent years, drug injection has been the leading cause of these infections. These infections are usually accompanied by a markedly elevated white blood cell count. Community-acquired MRSA has risen in prevalence in the past decade, with methicillin resistance now reported in 60 % of community isolates of *S. aureus*. MRSA also produces toxins that result in direct tissue injury.

In addition to these pathogens, several other bacteria have been associated with monomicrobial NSTIs. *Vibrio vulnificus* (saltwater injury) and *Aeromonas hydrophila* (freshwater injury) can produce monomicrobial NSTIs associated with water exposure but are more typically present in polymicrobial infections. Enterobacteriaceae, *Pseudomonas aeruginosa*, and *Yersinia enterocolitica* can also lead to monomicrobial NSTIs.

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_90, © Springer New York 2015

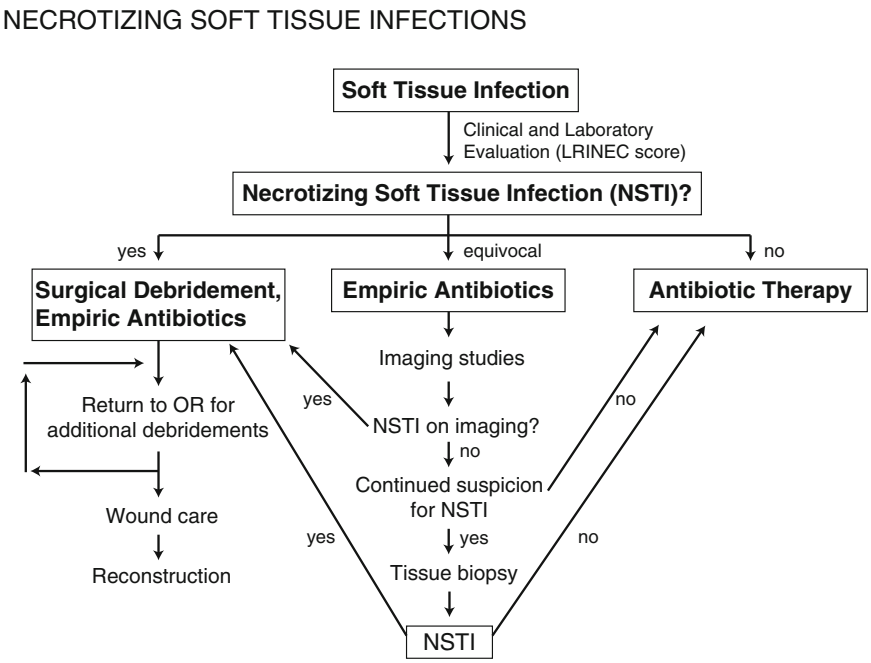

FIG. 90.1 Treatment algorithm for necrotizing soft tissue infections. *LRINEC* Laboratory Risk Indicator for Necrotizing Fasciitis

Clinical Manifestations and Diagnosis

Early suspicion and diagnosis of NSTIs is of paramount importance. A delay in diagnosis and treatment of more than 12 h is associated with increased mortality. High-risk populations for NSTIs include patients with a history of injection drug use, alcoholism, morbid obesity, diabetes mellitus, heart disease, renal failure, chronic skin infections, penetrating injuries, peripheral vascular disease, and immunosuppression.

Clinical manifestations include pain out of proportion to physical exam, erythema, induration, edema, bullae formation, skin necrosis with blue or black discoloration, crepitus, and thin gray ("dishwater") discharge. It is important to note that the infection may spread rapidly in the more susceptible subcutaneous tissues without producing equally noteworthy skin findings. Late clinical manifestations include signs of sepsis, such as fever, tachycardia, and hypotension.

Several studies have focused on using basic blood tests to predict the presence of NSTI (vs. a non-necrotizing soft tissue infection). The most commonly encountered laboratory abnormalities include leukocytosis, hyponatremia (<135 mmol/L), elevated blood urea nitrogen or creatinine, and elevated creatinine phosphokinase (CPK). The Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score is a model used to predict the presence of necrotizing fasciitis based on common laboratory parameters (see Table 90.1). Bacteremia may be present but is often found well after clinical presentation and is not a useful determinant of whether necrosis is present.

Although plain radiographs classically demonstrate gas in the soft tissues, this finding is present in only 50 % of cases, and its absence does not rule out NSTI. In equivocal cases, additional imaging with computed tomography (CT) or magnetic resonance imaging (MRI) may prove useful, but these studies should not delay operative intervention if there is high clinical suspicion.

Definitive diagnosis of NSTI is based on tissue pathology from surgical exploration. Bedside fascial biopsy has been

TABLE 90.1 The Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score. In this model, the minimum score is 0, and the maximum score is 13. A score of ≥6 should raise suspicion of necrotizing fasciitis; a score of ≥8 is strongly predictive of necrotizing fasciitis

Variable	Score
C-reactive protein (mg/L)	
<150	0
≥150	4
White blood cell count (per mm^3)	
<15	0
15–25	1
>25	2
Hemoglobin (g/dL)	
>13.5	0
11–13.5	1
<11	2
Sodium (mmol/L)	
≥135	0
<135	2
Creatinine	
≤141 μ(mu)mol/L (1.6 mg/dL)	0
>141 μ(mu)mol/L (1.6 mg/dL)	2
Glucose	
≤10 mmol/L (180 mg/dL)	0
>10 mmol/L (180 mg/dL)	1

described for equivocal cases but as with imaging studies should not delay prompt surgical exploration in cases of high suspicion. The presence of necrosis on the surgical biopsy confirms the diagnosis of NSTI initially made based on clinical suspicion and laboratory findings.

Treatment

Surgical Debridement

Surgical debridement is the cornerstone of treatment for NSTI; without debridement, NSTI mortality rates approach 100 %. Therefore, once the diagnosis of NSTI has been made, prompt, aggressive debridement of all necrotic tissue should be performed. The fascial planes should be explored, with the extent of debridement dependent upon the intraoperative findings; if necessary, amputation is performed. The goals of surgery are debridement of frank necrosis back to normal bleeding tissue and drainage of all fluid collections while sparing as much viable tissue as possible. Close attention is paid to hemostasis. NSTIs typically require several returns to the operating room until there is no further progression of necrosis. Wound care is managed initially with wet-to-dry dressings but may ultimately involve negative-pressure therapy once the infection is clearly resolving. A diverting colostomy may be required for perineal/perirectal wounds.

Antibiotic Therapy

Empiric administration of broad-spectrum antibiotics should begin as soon as NSTI is suspected. The antibiotics should cover the likely microbiology of the wound but typically consist of coverage for gram-positive, enteric gram-negative, and anaerobic organisms *as well as additional coverage for MRSA*. Regimens for polymicrobial infections should include imipenem-cilastatin, meropenem, piperacillin-tazobactam, ticarcillin-clavulanate, or tigecycline in combination with vancomycin or linezolid. For toxin-producing organisms of monomicrobial infection, clindamycin should be used, as its protein-synthesis inhibition is thought to reduce toxin production. Penicillin should be added if clostridial infection is suspected.

Adjuvant Therapies

Several adjuvant therapies have been proposed for NSTIs, but none has proven benefits for routine use. Hyperbaric oxygen (HBO) is known to be bactericidal for *C. perfringens* and reduces toxin production. Intravenous immunoglobulin (IVIG) can bind superantigens and has been used in the treatment of staphylococcal toxic shock syndrome; it likely reduces the toxicity of other toxin-producing organisms as well. Plasmapheresis and plasma exchange reduce the toxin and cytokine load present in severe sepsis caused by NSTIs. No adjuvant therapy should replace or delay surgical debridement.

91
Systemic Inflammatory Response Syndrome (SIRS) and Sepsis

Daniel Rinewalt and José M. Velasco

Introduction

The human body reacts to various insults with a variety of complex reactions. The **systemic inflammatory response syndrome (SIRS)**, first identified by Roger Bone, M.D., refers to a diffuse state of inflammation resulting from many possible etiologies, including, but not limited to: infection, drugs, toxins, adrenal insufficiency, trauma, burns, ischemia, and pancreatitis. Activation of the immune system causes lymphocytes and macrophages to release a variety of cytokines (IL-1, IL-6, IL-8, TNF alpha), enzymes, and vasoactive substances, whose function is to eradicate foreign organisms. Vast amplification of these same mediators may have several unwanted effects including rampant activation of the coagulation cascade, increased metabolism, and capillary permeability. When prolonged, this exaggerated immune system response constitutes the basis of SIRS.

While SIRS may result from infectious or noninfectious causes, **sepsis** by definition is a widespread inflammatory state secondary to a bacterial, fungal, or viral infectious agent. As a continuum of pathological inflammation, *sepsis can be understood as SIRS with a proven source of infection* (Fig. 91.1). A considerable cause of morbidity and mortality especially in intensive care unit (ICU) patients, sepsis can accompany urinary tract infections (UTIs) and pneumonia or be a complication of other disease processes such as trauma or cancer (e.g., skin infections in burn patients or colonic perforation from an obstructing tumor). **Severe sepsis** may be accompanied by organ dysfunction or signs of hypoperfusion. **Septic shock** is manifested by severe sepsis with hypotension, which may progress towards multiple organ dysfunction syndrome (MODS). Prompt diagnosis and treatment are essential in order to prevent its development and its high mortality rate.

Clinical Presentation

Patients with SIRS will commonly present with a combination of fever, tachycardia, tachypnea, and leukocytosis. Sepsis will usually have similar signs; however, it is important to note that hypothermia and neutropenia are also common. Invariably, sepsis will have evidence of infection via positive blood cultures, urine cultures, chest X-ray findings consistent with pneumonia, or the presence of a disease process known to cause systemic infection such as a perforated viscus or wet gangrene of an extremity.

Worsening sepsis progressing to grave conditions such as septic shock and multiple organ dysfunction syndrome will be accompanied by coagulopathy, metabolic acidosis, and increasing hemodynamic instability not responding to fluid resuscitation. Symptoms of organ dysfunction include acute respiratory distress syndrome (ARDS), encephalopathy, hyperbilirubinemia, oliguria, electrolyte abnormalities, and congestive heart failure.

Diagnosis

Guidelines for the diagnosis of SIRS, sepsis, severe sepsis, and septic shock have all been firmly established.

In order to make the diagnosis of SIRS, at least two of the following criteria must be met:

- Temperature greater than 38 °C or less than 35 °C
- Pulse greater than 90 beats per minute
- Respiratory rate greater than 20/min or $PaCO_2$ less than 32 mmHg
- White blood cell count greater than 12,000 or less than 4,000 mm^3

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_91, © Springer New York 2015

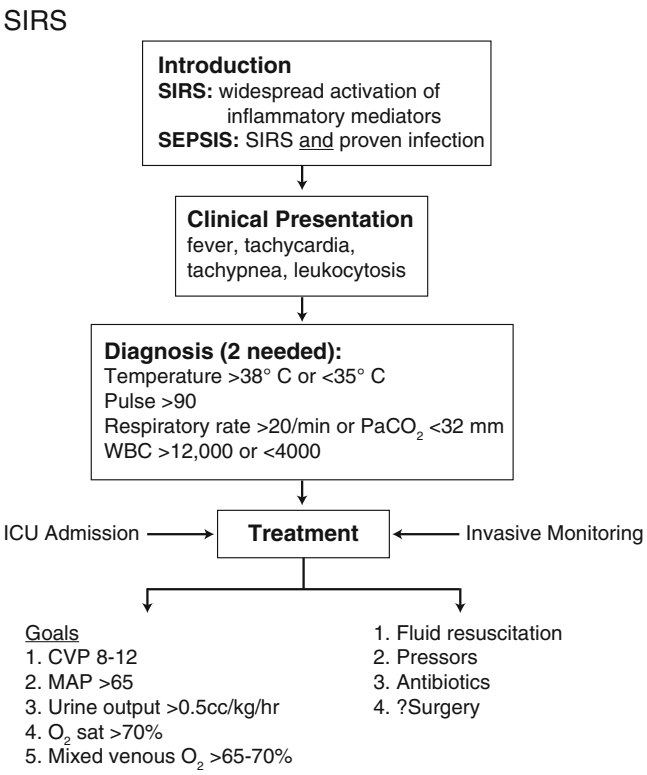

FIG. 91.1 Treatment algorithm for SIRS and sepsis. *CVP* central venous pressure, *MAP* mean arterial pressure

Criteria for the diagnosis of sepsis includes meeting the requirements for SIRS along with evidence of infection. Severe sepsis is diagnosed when a patient meets all criteria for sepsis and also has some degree of organ dysfunction or hypoperfusion. Diagnosis of septic shock consists of severe sepsis and hypotension, defined as a systolic blood pressure less than 90 mm of Hg or greater than 90 mm of Hg with the assistance of vasopressors. Other causes of hypotension, most notably heart failure, should be ruled out with bedside echocardiography.

Treatment

Initial treatment for SIRS and sepsis centers on early goal-directed resuscitation of the patient during the first 6 h after recognition. If the patient's condition worsens or progresses to severe sepsis or shock, expedient transfer to an intensive care unit should be followed by placement of central venous access and arterial lines for invasive blood pressure monitoring.

The Surviving Sepsis Campaign (SSC), a collaboration of multiple critical care organizations, has defined the following goals of resuscitation:

- Central venous pressure between 8 and 12 mm of Hg
- Mean arterial pressure (MAP) greater than 65 mm of Hg
- Urine output greater than 0.5 mL per kg per hour
- Central venous oxygen saturation greater than 70 %
- A mixed venous oxygen saturation greater than 65 %
- Normalization of lactate

In cases where hypotension persists in spite of aggressive resuscitation with administration of either **colloid or crystalloid fluids** (typically after 2 l of crystalloids, initially 30 mL/kg crystalloid for hypotension or lactate 4mmol/L), **vasopressors** should be started and titrated to achieve a MAP above 65 mm of Hg as detailed above. *Norepinephrine* (levophed) is the most effective initial vasopressor; however, if it fails to achieve an adequate blood pressure, epinephrine should be given. Epinephrine, phenylephrine, or vasopressin should not be administered as the initial vasopressor in septic shock. *Vasopressin* 0.03 units/min may be subsequently added to norepinephrine with anticipation of an effect equivalent to norepinephrine alone. Dobutamine inotropic therapy should be considered when echocardiography shows a low cardiac output despite fluid resuscitation and combined vasopressor therapy. Administration of **broad-spectrum antibiotics** should be given after blood cultures are obtained and within 1 h of the diagnosis of severe sepsis or shock.

If heart failure is suspected (patient history, fluid overload on chest radiograph, peripheral edema), an echocardiogram should be obtained. Appropriate imaging studies should be obtained as soon as possible, in order to promptly identify the source of infection. **Operative therapy** may include removal of foreign bodies (invasive catheters), tissue debridement, drainage of abscesses, and laparotomy. Antibiotic coverage should be de-escalated to the particular infectious agent, when identified, as soon as feasible.

Administration of stress-dose steroids can be considered in cases of hypotension not responding to fluids or vasopressors. Hydrocortisone at a dose of 200 mg per day is the preferred medication and should not be weaned until vasopressors are no longer needed. The SSC also recommends the use of blood products to keep the hemoglobin above 7 g/dL, and platelet counts above 5,000/mm^3, or greater than 50,000/mm^3 when invasive procedures are required. In cases where bleeding is less likely, recombinant activated protein C was considered in an attempt to reduce the production of cytokines and inhibit activation of the clotting cascade, specifically in adult patients with an Acute Physiology and Chronic Health Evaluation (APACHE) score greater than 25 or multiorgan failure. This medication is no longer available nor considered for use.

Acute lung injury and acute respiratory distress syndrome are best managed by the judicious use of fluids, low tidal volume, limitation of inspiratory plateau pressure, and application of positive end-expiratory pressure. Other maneuvers include: head-of-bed elevation in mechanically ventilated patients, avoiding routine insertion of pulmonary artery catheters, protocols for weaning and sedation, and avoidance of neuromuscular blockers. Institution of glycemic control when two consecutive blood glucose levels are >180 mg/dL is recommended. This protocolized approach should target an upper blood glucose ≤180 mg/dL rather than an upper target blood glucose ≤150 mg/dL. The use of stress ulcer and DVT prophylaxis is also strongly recommended therapeutic maneuvers.

ERRATUM TO

Surgical Hypertension: Evaluation and Treatment

Leon Boudourakis and Kaare J. Weber

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1_28, © Springer New York 2015

DOI 10.1007/978-1-4939-1565-1_92

The incorrect affiliation was published for the corresponding author of Chapter 28 (Surgical Hypertension: Evaluation and Treatment) Leon Boudourakis. Here is the correct affiliation:

Leon Dimitrios Boudourakis
Assistant Professor of Surgery
Trauma & Critical Care
SUNY Downstate Medical Center
Kings County Hospital

The online version of the original chapter can be found at
http://dx.doi.org/10.1007/978-1-4939-1565-1_28

Index

T.J. Saclarides et al. (eds.), *Common Surgical Diseases: An Algorithmic Approach to Problem Solving*,
DOI 10.1007/978-1-4939-1565-1, © Springer New York 2015