TEXAS CHILDREN'S HOSPITAL
EVIDENCE-BASED OUTCOMES CENTER
Acute Decompensated Heart Failure (ADHF)
Evidence-Based Guideline

Definition: Acute decompensated heart failure (ADHF) is defined as either new-onset heart failure or decompensation of chronic heart failure with symptoms that warrant hospitalization. ADHF may be caused by cardiac dysfunction as a result of myocardial muscle dysfunction or loss characterized by left ventricular dilation or hypertrophy. Dysfunction may manifest as primarily systolic, diastolic or mixed. Clinical symptoms can vary substantially during the course of the disease process and may not correlate with changes in underlying cardiac function. ADHF is progressive and often fatal, but myocardial dysfunction and remodeling may improve spontaneously or as a result of therapy. In physiologic terms, ADHF is a clinical syndrome characterized by inadequate peripheral oxygen delivery, at rest or during stress, caused by cardiac dysfunction.

Epidemiology: While the incidence and prevalence of pediatric heart failure is hard to measure, there were an estimated 13,892 admissions (95% CI 11, 528-16,256) pediatric heart failure-related admissions in 2006. [(1)] This population of children has a high unadjusted mortality rate at 7.4% when compared to hospitalized children without heart failure (0.4%). and a mean hospital duration of 19.4 days. [(1)]

Etiology: ADHF can result from congenital cardiac malformations or from a structurally normal heart. In congenital cardiac malformations, the patient may have volume overload secondary to left-to-right shunting (from ventricular septal defect or patent ductus arteriosis) or an atrioventricular or semilunar valve insufficiency, pressure overload from either a left-sided or right-sided obstruction, or complex congenital heart disease such as single ventricle or transposition of the great arteries. In the structurally normal heart, primary cardiomyopathies can be dilated, hypertrophic, or restrictive; secondary cardiomyopathies can be due to arrhythmogenic, ischemic, toxic, infiltrative, or infectious agents. [(2)]

Inclusion Criteria
- Age birth - 21 years
- Chronic heart failure with acute decompensation
- New onset acute decompensated heart failure
- Cardiogenic shock

Exclusion Criteria
- Adults (>21 years old)
- Neonates post-menstrual age <35 weeks
- Infants with a potential need for prostaglandin infusion
- Infants receiving prostaglandins

Differential Diagnosis
- Sepsis/Shock
- Respiratory Failure
- Gastroenteritis
- Hepatitis
- Primary lung disease- asthma, bronchiolitis, pneumonia
- Renal failure and volume overload

Diagnostic Evaluation
History: Assess for

Relevant medical problems and description of symptoms (intermittent or constant, progressive, prior episodes, duration, aggravating or alleviating features). Age at presentation should drive history and physical examination.

Respiratory*: Increased work of breathing (WOB), tachypnea, or dyspnea at rest or with minimal activity
Cardiac: Congenital heart defects (CHDs) with or without operative intervention. May include: ductal dependent defects for systemic or pulmonary perfusion or CHDs with increasing left-to-right shunting of blood flow; use of routine medications
Nutritional: Weight gain or loss;
Gastrointestinal*: Nausea and/or vomiting
Genitourinary: Decreased urine output
Activity: Sleeping patterns (not sleeping well or sleeping too much)
**Presentation with respiratory and gastrointestinal symptoms that are disproportionate to HPI may be indicative of acute heart failure syndromes*.

Neonates/Infants:
Irritability
Feeding (generally poor feeding, tires easily, extended time to complete feeding, tachypnea with feeds, diaphoresis)
Perinatal asphyxia, incessant tachyarrhythmias, neonatal myocarditis, or severe anemia.

Children/Adolescents:
Exercise intolerance
Easy fatigability
Orthopnea and/or paroxysmal nocturnal dyspnea†
Edema as a result of fluid overload†
†These signs and symptoms are rare but may be seen in school-age children and adolescents.

Past medical history:
Age at presentation should guide assessment of medical history. All histories should include:
- Viral history
- Family history (cardiomyopathy)
- Prior heart transplantation
- Presence of chronic heart failure
- Existing CHDs
- Medical or surgical interventions for CHD
- Rheumatic heart disease
- Hypothyroidism
- Kawasaki disease
- Cancer/Chemotherapy (e.g., anthracyclene)

Physical Examination:
The severity of ADHF is based on overall clinical appearance and behavior, including the child's alertness, respiratory effort, and ability to take oral fluids/nutrition.

A complete physical exam should be performed. A combination of clinical findings is most predictive when determining ADHF. The most common presenting signs and symptoms of ADHF include: [2]
- Dyspnea or increased work of breathing (WOB)

- Tachypnea
- Wheezing (especially new onset)
- Hepatomegaly
- Nausea or vomiting
- Abdominal distension/pain
- Fatigue or decreased activity level
- Peripheral edema‡

‡Absence of edema does not preclude a diagnosis of ADHF.

Clinical management will vary based upon the presentation. (See Table 1).

Table 1. Presentations of ADHF [2]

Warm & Dry	Warm & Wet
Represents asymptomatic ventricular dysfunction (normal filling pressures and adequate perfusion). The primary focus is on the prevention of disease progression and decompensation.	The most common presentation and can be characterized by elevated filling pressures and pulmonary edema with adequate perfusion.
Cold & Wet	**Cold & Dry**
Characterized by elevated filling pressures and poor perfusion. These patients often require intensive care management.	A dire condition best described as the presence of normal filling pressures with poor perfusion. These patients require aggressive treatment to minimize the workload of the myocardium.

Other signs and symptoms include: tachycardia, gallop heart rhythm, cool skin, cough and hypertension. Cardiac murmurs; single, second heart sounds; clicks; pulses and decreased oxygen saturation are indicative of ADHF.
Neonates and infants present with irritability or lethargy and signs and symptoms of pain especially during feeding.

Laboratory Tests:
Diagnostic and laboratory testing are useful tools that provide additional information for diagnosing ADHF.

Initial diagnostic testing:
Chest X-ray to assess for cardiomegaly (a cardiothoracic ratio of >0.55 in infants and >0.5 in children)
B-type natriuretic peptide levels (BNP)
Complete Blood Count (CBC)
Chemistries
A capillary or arterial blood gas may be included in the initial evaluation if shock, impending respiratory failure or s/sx of hypoxia are present.
Electrocardiography (ECG) to assess for arrhythmias; findings may include atrial enlargement, ventricular hypertrophy, strain and changes in ST-segment or T-wave morphology. ECG is most useful in cases of ADHF originating from myocarditis, anomalous left coronary artery from the pulmonary artery (ALCAPA), tachyarrhythmia-induced cardiomyopathy or restrictive cardiomyopathy.
Consultation with a cardiologist is recommended if initial diagnostic testing is indicative of ADHF.

Subsequent diagnostic testing:
Echocardiography (ECHO) is the most precise way to quickly evaluate cardiac function. [3] Cardiac anatomy is easily assessed and estimations of gradients, shunting and cardiac output can be made.

Critical Points of Evidence

Evidence Supports

- Perform a thorough history and physical examination combined with supportive evidence from chest x-ray imaging, ECG, echocardiography, and laboratory evaluations, including BNP. (4-7) – Strong recommendation, very low quality evidence.
- Perform chest x-ray imaging, EKG, and BNP if a clinician has a suspicion for cardiac disease. (4, 8-24) – Strong recommendation, very low quality evidence
- Administer a loop diuretic bolus dose without delay to patients who present with signs and symptoms of congestion. Reassess fluid status before administering additional doses. (25-32) – Strong recommendation, very low quality evidence.
- Administer beta-blockers to patients prior to discharge in those who have biventricular anatomy with reduced ejection fraction. This should be started at a slow dose and should be up-titrated to a maximum tolerated safe dose. If a patient presents with acute heart failure who is already taking beta-blockers, continue the beta-blocker treatment unless they have second or third degree atrioventricular block or shock. (33-37) – Strong recommendation, low quality evidence
- Administer ACE inhibitors in the treatment of left ventricular dysfunction. This should be started at a low dose and should be up-titrated to a maximum tolerated safe dose. (34, 37) – Strong recommendation, low quality evidence
- Use milrinone in children who present with heart failure due to reduced cardiac output with end-organ dysfunction as a rescue strategy. (38-41) – Strong recommendation, low quality evidence.
- Consider using non-invasive ventilation strategies in patients with heart failure, pulmonary edema, and respiratory insufficiency. Identify those patients who are non-responders as patients exhibiting no response (e.g. HR, RR) within one hour of therapy. (42-48) – Weak recommendation, low quality evidence

Evidence Lacking/Inconclusive

- Initiate 13-blocker therapy as soon as possible after stabilization and discontinuation of inotropic support. 13-blocker therapy should be initiated prior to discharge. (35,49) – Consensus recommendation
- Using cerebral near-infrared spectroscopy (NIRS) as accurate as BNP in diagnosing ADHF.
- Using milrinone to improve myocardial performance instead of DOPamine in neonates with ADHF. (50-52)
- Use of Intermacs criteria to help determine the need for temporary circulatory support. (53-55)

Evidence Against

- Using BNP for diagnosing heart failure in neonates. (19,56-61) – Strong recommendation, low quality evidence
- Routine use of beta-blockers in single ventricle anatomy. (33-37) – Strong recommendation, low quality evidence

- Routine administration of calcium chloride to improve myocardial performance in patients who are beyond the neonatal period. (62) – Strong recommendation, very low quality evidence
- Use of sodium bicarbonate in children with acidosis secondary to acute decompensated heart failure. (63-65) – Strong recommendation, very low quality evidence

Condition-Specific Elements of Clinical Management

General: Signs and symptoms of acute decompensated heart failure can often mimic those of other common pediatric diseases such as gastroenteritis.

Admission Criteria

To NICU/PICU/CvICU

Impending respiratory failure
Respiratory distress
Unstable arrhythmias
Atrio-ventricular heart block
Requires continuous vasodilator, natriuretic peptide, or inotropic support
Complex congenital heart defects
Requires IV narcotics for comfort

To Cardiac Stepdown

Acute exacerbation of heart failure in the presence of chronic heart failure that improves with initial management

Transfer Criteria (from ICU to Stepdown)*

*Patients admitted to Level III NICU may transfer to Level II or a Stepdown bed based on availability
Tolerating enteral nutrition
No longer requires continuous vasodilator, natriuretic peptide or inotropic support OR transition to stable continuous dosing
Successfully weaning from oxygen requirements
No arrhythmias requiring treatment in past 24 hours
Improved or stable ventricular heart function

Discharge Criteria

Patient in stable, compensated state on oral medication

Consults/Referrals

Consultation with a cardiologist is recommended for all patients presenting to the EC with ADHF.

Measures:

Process

- Time to first diuretic dose
- Time to inpatient disposition (from EC to admission)
- Length of stay
- Incidence of BNPs ordered in EC
- Incidence of Chest X-rays ordered in EC
- Incidence of ECHOs ordered in EC for newly diagnosed AHFS
- Incidence of vasoactive agent use when contraindicated by clinical picture
- Appropriate use of captopril and enalapril related to age
- Incidence of calcium use
- Incidence of nesiritide use
- Incidence of diuretic use

Outcome

- Mortality related to nesiritide use
- Incidence of development of renal failure/insufficiency
- (creatinine levels)
- Incidence of arrhythmias
- Incidence of discharge without ACE inhibitor and/or β blocker
- Incidence of readmission to hospital within 30 days of discharge
- Incidence of presentation to EC within 14 days of discharge

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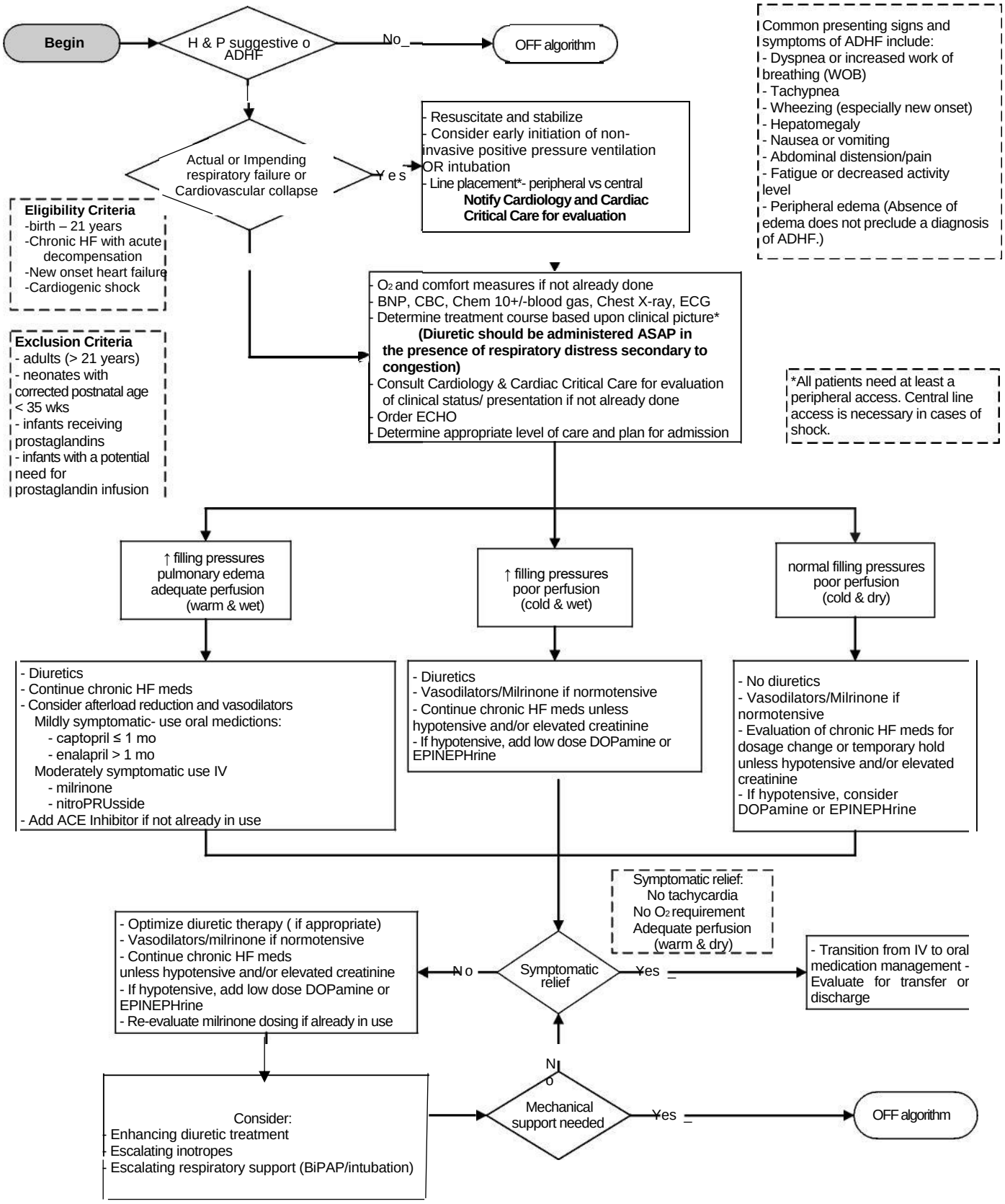
TCH Evidence-Based Outcomes Center
Clinical Algorithm for Acute Decompensated Heart Failure (ADHF)



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Clinical Standards Preparation

This clinical standard was prepared by the Evidence-Based Outcomes Center (EBOC) team in collaboration with content experts at Texas Children's Hospital. Development of this clinical standard supports the TCH Quality and Patient Safety Program initiative to promote clinical standards and outcomes that build a culture of quality and safety within the organization.

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Development Process

This clinical standard was developed using the process outlined in the EBOC Manual. The literature appraisal documents the following steps:

1. Review Preparation
   - PICO questions established
   - Evidence search confirmed with content experts

2. Review of Existing Internal and External Guidelines
   - International Society for Heart and Lung Transplantation (2014) "Guidelines for management of pediatric heart failure," Canadian Cardiovascular Society (2013) "Presentation, Diagnosis, and Medical Management of Heart Failure in Children," American College of Cardiology Foundation/American Heart Association (2013) "ACCF/AHA Guideline for the Management of Heart Failure," National Institute for Health and Care Excellence (2014) "Acute Heart Failure: Diagnosing and managing acute heart failure in adults," European Society of Cardiology (2012) "ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure," and the Heart Failure Society of America (2010) "Heart Failure Society Comprehensive Heart Failure Practice Guideline."

   0. Literature Review of Relevant Evidence
      - Searched: PubMed, CINAHL, Cochrane, Guideline Clearinghouse

   1. Critically Analyze the Evidence
      - 9 meta-analyses, 7 randomized controlled trials, and 47 nonrandomized studies

   2. Summarize the Evidence
      - Materials used in the development of the guideline, evidence summary, and order sets are maintained in an Acute Decompensated Heart Failure evidence-based review manual within EBOC.

3. **Evaluating the Quality of the Evidence**

Published clinical guidelines were evaluated for this review using the **AGREE II** criteria. The summary of these guidelines are included in the literature appraisal. AGREE II criteria evaluate Guideline Scope and Purpose, Stakeholder Involvement, Rigor of Development, Clarity and Presentation, Applicability, and Editorial Independence using a 4-point Likert scale. The higher the score, the more comprehensive the guideline.

This clinical standard specifically summarizes the evidence *in support of* or *against* specific interventions and identifies where evidence is *lacking/inconclusive*. The following categories describe how research findings provide support for treatment interventions. ***"Evidence Supports"*** provides clear evidence that the benefits of the intervention exceed harm.

"Evidence Against" provides clear evidence that the intervention is likely to be ineffective or that it is harmful.

"Evidence Lacking/Inconclusive" indicates there is currently insufficient data or inadequate data to support or refute a specific intervention.

The **GRADE** criteria were utilized to evaluate the body of evidence used to make practice recommendations. The table below defines how the quality of the evidence is rated and how a strong versus weak recommendation is established. The literature appraisal reflects the critical points of evidence.

Recommendation	
STRONG	Desirable effects clearly outweigh undesirable effects or vice versa
WEAK	Desirable effects closely balanced with undesirable effects
Quality	**Type of Evidence**
High	Consistent evidence from well-performed RCTs or exceptionally strong evidence from unbiased observational studies
Moderate	Evidence from RCTs with important limitations (e.g., inconsistent results, methodological flaws, indirect evidence, or imprecise results) or unusually strong evidence from unbiased observational studies
Low	Evidence for at least 1 critical outcome from observational studies, RCTs with serious flaws or indirect evidence
Very Low	Evidence for at least 1 critical outcome from unsystematic clinical observations or very indirect evidence

Recommendations

Practice recommendations were directed by the existing evidence and consensus amongst the content experts. Patient and family preferences were included when possible. The Content Expert Team and EBOC team remain aware of the controversies in the diagnosis/management of acute decompensated heart failure in infants, children, and young adults. When evidence is lacking, options in care are provided in the clinical standard and the accompanying order sets (if applicable).

Approval Process

Clinical standards are reviewed and approved by hospital committees as deemed appropriate for its intended use. Clinical standards are reviewed as necessary within EBOC at Texas Children's Hospital. Content Expert Teams are involved with every review and update.

Disclaimer

Practice recommendations are based upon the evidence available at the time the clinical standard was developed. Clinical standards (guidelines, summaries, or pathways) do not set out the standard of care, and are not intended to be used to dictate a course of care. Each physician/practitioner must use his or her independent judgment in the management of any specific patient and is responsible, in consultation with the patient and/or the patient family, to make the ultimate judgment regarding care.

Version History

Date	Action	Comments
Sep 2009	Originally completed	
Apr 2017	Updated	
Mar 2023	Reaffirmed	