STANDARD TREATMENT WORKFLOW (STW)

ACUTE AORTIC SYNDROME

Shiv Choudhary[1], Gautam Ganguli[2], Sai Chandran BV[3], Dhiren Shah[4], T Sunder[5], Lokeshwar Rao Sajja[6], Shantanu Pande[7], Ambuj Roy[8], Arun Kumar Yadav[9]

[1]All India Institute of Medical Sciences Delhi; [2]Armed Forces Medical College, Pune Maharashtra; [3]Jawaharlal Institute of Postgraduate Medical Education and Research Pondicherry; [4]Ahmedabad; [5]Apollo Hospital, Chennai; [6]Telangana; [7]Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow; [8]All India Institute Of Medical Sciences Delhi; [9]Armed Forces Medical College, Pune Maharashtra

CORRESPONDING AUTHOR

Shiv Choudhary, All India Institute of Medical Sciences Delhi

Email: shivchoudhary@hotmail.com

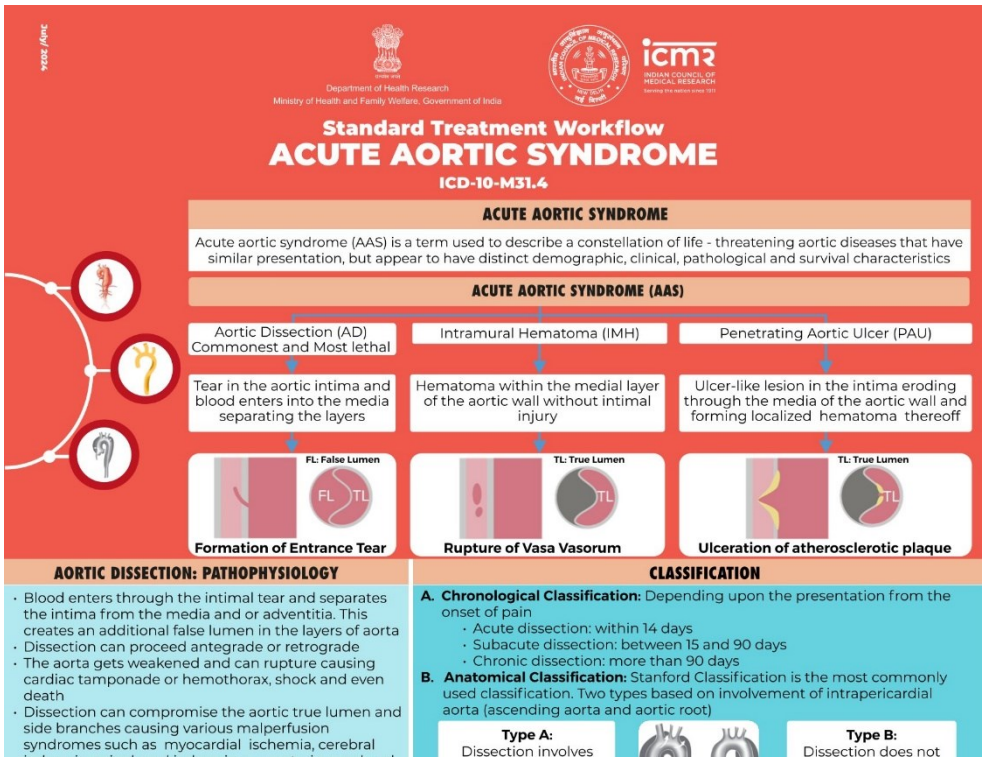
CITATION

Choudhary S, Ganguli G, Chandran SBV, Shah D, Sunder T, Sajja LR, Pande S, Roy A, Yadav AK. ACUTE AORTIC SYNDROME. Journal of the Epidemiology Foundation of India. 2024;2(2Suppl):S251-S254.

DOI: https://doi.org/10.56450/JEFI.2024.v2i2Suppl.001

DISCLAIMER

This article/STW, was originally published by Indian Council of Medical Research (ICMR) under Standard Treatment Workflow. The reprinting of this article in Journal of the Epidemiology Foundation of India (JEFI) is done with the permission of ICMR. The content of this article is presented as it was published, with no modifications or alterations. The views and opinions expressed in the article are those of the authors and do not necessarily reflect the official policy or position of JEFI or its editorial board. This initiative of JEFI to reprint STW is to disseminate these workflows among Health Care Professionals for wider adoption and guiding path for Patient Care.



July 2024

Department of Health Research
Ministry of Health and Family Welfare, Government of India

icmr INDIAN COUNCIL OF MEDICAL RESEARCH

Standard Treatment Workflow
ACUTE AORTIC SYNDROME

ICD-10-M31.4

ACUTE AORTIC SYNDROME

Acute aortic syndrome (AAS) is a term used to describe a constellation of life - threatening aortic diseases that have similar presentation, but appear to have distinct demographic, clinical, pathological and survival characteristics

ACUTE AORTIC SYNDROME (AAS)

Aortic Dissection (AD) Commonest and Most lethal	Intramural Hematoma (IMH)	Penetrating Aortic Ulcer (PAU)
Tear in the aortic intima and blood enters into the media separating the layers	Hematoma within the medial layer of the aortic wall without intimal injury	Ulcer-like lesion in the intima eroding through the media of the aortic wall and forming localized hematoma thereoff

AORTIC DISSECTION: PATHOPHYSIOLOGY

- Blood enters through the intimal tear and separates the intima from the media and or adventitia. This creates an additional false lumen in the layers of aorta
- Dissection can proceed antegrade or retrograde
- The aorta gets weakened and can rupture causing cardiac tamponade or hemothorax, shock and even death
- Dissection can compromise the aortic true lumen and side branches causing various malperfusion syndromes such as myocardial ischemia, cerebral ischemia, spinal cord ischemia, mesenteric, renal and limb ischemia
- If the dissection involves aortic root it may cause acute aortic valve insufficiency
- If a patient survives acute episode, the false lumen may either get thrombosed or may dilate aneurysmally

Two factors are primarily responsible for pathogenesis of dissection

- Higher wall stress due to hypertension or dilatation of aorta
- Inherently weak/degenerated aortic media

Predisposing factors

- The commonest risk factor is hypertension.
- Genetically mediated aortopathies like
  - Marfan's syndrome
  - Vascular Ehlers-Danlos syndrome
  - Bicuspid aortic valve
  - Familial aortic dissection

Trauma and iatrogenic injury are also emerging as important causes of aortic dissection

CLASSIFICATION

A. Chronological Classification: Depending upon the presentation from the onset of pain
- Acute dissection: within 14 days
- Subacute dissection: between 15 and 90 days
- Chronic dissection: more than 90 days

B. Anatomical Classification: Stanford Classification is the most commonly used classification. Two types based on involvement of intrapericardial aorta (ascending aorta and aortic root)

Type A: Dissection involves the intrapericardial aorta regardless of the site of intimal tear. It is also known as Proximal dissection

Type B: Dissection does not involve the intrapericardial aorta. It is also known as Distal dissection

CLINICAL PRESENTATION AND COMPLICATIONS

- Abrupt onset of severe chest and/or back pain is the most typical feature.
- The pain is sharp, ripping, tearing, knife-like, and typically different from other causes of chest pain.
- Anterior chest pain is more common in acute Type A dissection, whereas back or abdominal pain is more common with Type B dissection.
- The pain may migrate from its point of origin to other sites, following the dissection path
- Other manifestations may include
  - Shock
  - Syncope
  - Neurological deficit including stroke and paraplegia
  - Acute congestive heart failure
  - Myocardial ischemia
  - Lower extremity ischemia
  - Renal and visceral ischemia
  - Sudden death

IMPORTANT COMPLICATIONS OF DISSECTION, CAUSATIVE MECHANISMS, AND PRESENTATIONS

COMPLICATION	CAUSATIVE MECHANISM	PRESENTATION
Aortic Regurgitation	Loss of support and distortion of aortic valve	Diastolic murmur, Severe acute regurgitation can cause heart failure and pulmonary edema
Myocardial Ischemia	Dissection process involves one or more coronary ostia	Chest pain and ECG suggestive of myocardial infarction. Cardiac enzymes may be raised
Congestive heart failure	Acute severe aortic regurgitation, extensive myocardial ischemia	Hypotension, features of low cardiac output
Cardiac Tamponade	Leak/rupture of aorta in pericardial cavity	Raised JVP, muffled heart sounds, hypotension
Pleural effusion	Leak/rupture of aorta in pleural cavity	Diminished breath sounds. Diagnosed on X-ray, Ultrasound, CT Scan
Hypotension and Shock	Hypotension and shock may result from aortic rupture, acute severe aortic regurgitation, extensive myocardial ischemia, cardiac tamponade	Hypotension, features of low cardiac output
Neurological Complication	Involvement of arch vessels or intercostal arteries, low output syndrome, pericardial tamponade, hypoxia.	Confusion/agitation, Syncope, Stroke, Coma, Paraplegia/paraparesis
Mesenteric malperfusion	Involvement of visceral arteries, Compression of true lumen	Hypotension, abdominal distension, absent bowel sounds, shock, acidosis
Renal failure	Involvement of renal arteries, Compression of true lumen, low output syndrome	Abnormal renal function tests and decreased urine output
Acute Lower Limb Ischemia	Involvement of iliac arteries, Compression of true lumen	Pain in lower limbs, absent pulses, cold and pale limbs, gangrene
Inflammatory Syndrome	Inflammatory response to blood /thrombus in the media	Fever, Leucocytosis

MYOCARDIAL INFARCTION VS ACUTE TYPE A DISSECTION

- Clinically, a dissection-related coronary malperfusion may present with ECG changes of primary myocardial ischemia or infarction. This may increase the likelihood of misdiagnosis and inappropriate therapeutic intervention
- Before starting the treatment algorithm for myocardial ischemia, it is important to differentiate between a primary coronary event and coronary malperfusion secondary to dissection process
- In dissection, pain is usually abrupt, piercing, and well localized. Presence of diastolic murmur of aortic regurgitation, unequal pulses, limb ischemia or neurological complications favor the diagnosis of acute dissection
- If dissection is diagnosed with myocardial ischemia it should be managed on the line of dissection and thrombolytic therapy should NOT be initiated

DIAGNOSTIC IMAGING IN ACUTE AORTIC DISSECTION

- The main purpose of imaging is the comprehensive assessment of the entire aorta and its branches with primary goals of confirmation of diagnosis, classification of dissection, assessment of branch vessels, and identification of complications (aortic rupture, aortic regurgitation, and malperfusion)
- Besides these, imaging is also needed to assess myocardial and valvular function, pulmonary parenchyma, and state of kidneys

IMAGING AND DIAGNOSTIC ALGORITHM

- Combination of ECG, Chest X-ray, Echocardiography and ECG gated CT angiography provides desired information. If clinical examination or CT angiography suggests involvement of innominate artery, common carotid arteries, subclavian arteries, and iliac arteries, an additional vascular ultrasound examination is performed. Conventional catheter based coronary angiogram and aortogram are not needed in diagnostic work up of acute dissection
- In a stable patient with low index of suspicion of dissection, D-dimers can be assessed. Normal levels of D-dimers virtually rule out dissection. However, IMH and PAU may still be present. In dissection, the level of D-dimers is immediately very high, compared with other disorders in which the D-dimers level increases gradually

If Acute Aortic Syndrome is suspected kindly refer the patient to higher center

July 2024

Department of Health Research
Ministry of Health and Family Welfare, Government of India

icmr INDIAN COUNCIL OF MEDICAL RESEARCH

Standard Treatment Workflow
ACUTE AORTIC SYNDROME
(Continued)

DIAGNOSTIC METHODS AND OUTPUT

Diagnostic method	Output
Chest X-ray	Any evidence of leak: widened mediastinum, pleural collection
CT angiography	Confirmation of diagnosis of dissection, Anatomical extent of dissection, Localization of entry and re-entry tears, Aortic rupture, State of arch vessels, coronary arteries, visceral arteries, renal arteries, and ilio-femoral arteries, State of lungs and kidneys
Transthoracic Echocardiography	Evaluation of aortic valve, Pericardial effusion, Evidence of tamponade, Myocardial function, Mitral and Tricuspid valve function
ECG	Evidence of myocardial ischemia
Vascular Ultrasound	State of axillary, carotid and femoral arteries

CT Angiography showing Type A Aortic Dissection

CT Angiography showing Type B Aortic Dissection

TREATMENT OF ACUTE DISSECTION

- Acute Type A dissection is a surgical emergency. Acute Type B dissection is mostly subjected to medical treatment unless complicated. Irrespective of surgical or endovascular intervention, medical therapy to control blood pressure and pain is essential

Initial Medical Therapy

- Initial management of dissection is directed at pain control and limiting the propagation of dissection by reducing the aortic wall stress. Aortic wall stress is affected by the velocity of ventricular contraction (dP/dt), the rate of ventricular contraction and blood pressure
- Initial medical treatment with beta blockers controls these 3 parameters by reducing heart rate and blood pressure to the lowest amounts that will still maintain adequate end-organ perfusion. Reasonable initial targets are a heart rate nearly 60/minute and a systolic blood pressure between 100 and 120 mm Hg
- In patients with severe aortic regurgitation, it is important to maintain a mean arterial pressure between 70-80 mm of Hg to ensure adequate end-organ perfusion. In presence of significant aortic regurgitation target heart rate is kept near 80/minute as lowering the heart rate furthur may prolong diastole and aggravate regurgitation

Dissection patients need management at specialized centers. Hence, after initiation of medical therapy, patient should be referred to specialized centre as soon as possible

ANTIHYPERTENSIVE DRUGS IN MANAGEMENT OF ACUTE DISSECTION

Group	Drugs	Dose	Special precaution/contraindication
β-Blockers (one of these to be used)	Labetalol	Loading: 20 mg IV in 2 minutes Maintenance: 1-2 mg/minute	Hypersensitivity, Severe asthma, Heart block, Uncompensated heart failure Severe chronic obstructive pulmonary disease, Severe Aortic Regurgitation (avoid extreme bradycardia)
	Esmolol	Loading : 250-500 µg/kg in one minute Maintenance: 50-100 µg/kg/min (maximum upto 300 µg/kg/min)	
	Metoprolol	Loading dose: 5mg IV over 2min, up to 3 doses; Maintenance dose: 3-5 mg every hour	
Calcium channel blockers (in addition to β blocker)	Diltiazem	Loading dose: 0.25 mg/kg over 2 to 5 min, Maintenance dose: 5mg/h IV infusion	Hypersensitivity, Atrioventricular block, Sick sinus syndrome, Ventricular dysfunction Pulmonary congestion
	Nicardipine	2.5-15 mg/hour IV infusion	
Vasodilator (in addition to β blocker)	Nitroprusside	0.3-0.5 µg/kg/min (Maximum upto 10 µg/kg/min)	Hypersensitivity

FLOW CHART FOR MEDICAL MANAGEMENT

Targets

Systolic BP 100-120 mm Hg
Mean arterial pressure 70mm Hg in significant aortic regurgitation
Heart Rate 60-70 beats/min
Adequate pain control

Initiation

Control of BP and Heart Rate: Start with Beta blocker
Pain Relief: IV Morphine 2-10 mg or Fentanyl 10- 50mcg

Targets achieved?

- Yes → Continue same
- No → Both Systolic BP and Heart Rate are high → Add Calcium Channel blocker
- No → Heart Rate controlled Systolic BP high → Add Vasodilator (Nitroprusside)

TYPE B DISSECTION REQUIRING INTERVENTION

Complications such as

- Unremitting pain
- Persistent, un-controlled hypertension
- Hemodynamic instability
- Rupture/Impending rupture/Leak
- Rapid Aortic Expansion
- Malperfusion
- Retrograde Ascending aortic dissection
- Aneurysmal dilatation (>5.5 cm)

SURGERY FOR ACUTE TYPE A DISSECTION

- Surgery is life - saving procedure in case of acute Type A dissection
- Type A dissection in itself is an indication for surgery
- The exceptions:
  - Extremely old and moribund patients with significant co-morbidities
  - Deeply comatose patients
  - Patients with advanced features of mesenteric ischemia, gangrene, and sepsis

Type A dissection is a surgical emergency and the ultimate aim is to save life

- This primary objective can be achieved by replacing the ascending aorta and excision of the primary tear
- Other objectives include correction of aortic regurgitation, treatment of coronary ostial dissection, restoration of distal true lumen and correction of malperfusion.

MANAGEMENT OF TYPE B DISSECTION

- Gold standard in management of Type B dissection
  - Optimal medical therapy
  - Control of blood pressure and pain
- Surgery or endovascular repair is reserved for complicated or high risk acute Type B dissections
- Surgery aims at resection of the primary intimal tear and ruptured/aneurysmal aorta
- **Thoracic Endo-vascular Aortic Repair (TEVAR)** aims at
  - Covering the primary intimal tear
  - Expansion of the true lumen
  - Thrombosis/remodeling of false lumen
  - Correction of malperfusion

MANAGEMENT OF IMH AND PAU

- The diagnosis of IMH and PAU is radiological and management is similar to acute aortic dissection

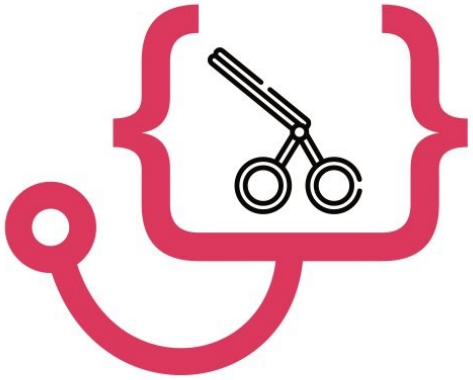
ABBREVATIONS

AAS: Acute Aortic Syndrome
AD: Aortic Dissection
CT: Computed Tomography
FL: False Lumen
IMH: Intramural Hematoma
JVP: Jugular Venous Pressure
PAU: Penetrating Aortic Ulcer
TL: True Lumen

REFERENCES

1. Choudhary SK, Bhoje A. Adult cardiac surgery. In: Prabhakaran D, Raman KK, Naik N, editors. Tandon's textbook of Cardiology. 1st edition. Walters Kluver (India); 2019: p679-727
2. Writing Committee Members, Isselbacher EM, Preventza O, Hamilton Black III J, Augoustides JG, Beck AW, Bolen MA, Braverman AC, Bray BE, Brown-Zimmerman MM, Chen EP. 2022 ACC/AHA guideline for the diagnosis and management of aortic disease: a report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines. Journal of the American College of Cardiology. 2022 Dec 13;80(24):e223-393

HIGH INDEX OF SUSPICION AT THE EARLIEST IS LIFE SAVING



STANDARD
TREATMENT
WORKFLOWS
of India