ACUTE THORACIC AORTIC DISSECTION

BRADFORD L. WALTERS, MD, FACEP
WILLIAM BEAUMONT HOSPITAL
DEPARTMENT OF EMERGENCY MEDICINE

DISCLAIMERS
• No conflicts of interest at all.
• Life Goal – to be the person my dog, Winston Churchill, thinks I am.

THE PROBLEM
• Few clinical entities make me sweat, wakes me up in the middle of the night, and haunts my dreams than an acute thoracic aortic dissection (ATAD).
• Frankly it is only a matter of time, seeing enough patients, and some bad luck (for you and the patient) before you miss an ATAD.
• So, it is worth exploring this problem to see if there is something in the literature that can help.

WHY IS THIS IMPORTANT
• Acute thoracic aortic dissection is highly lethal – mortality rates of 27% even under optimal conditions (in-patient) are seen.
• Often treatment in the ED are sub-optimal or encounter delays in definitive treatment as patients have to undergo testing and often transfer to another facility.
• There is a low incidence of the disease – 3.5 cases/100,000 population but expected to increase with our aging population.
• ATAD can be difficult clinically to diagnose, classic presentations are not as common as we would like, even silent presentations occur.
• The work-up most often does not find a dissection when it is suspected with only 2.7% of CT scans for ATAD are positive – that’s a very low rate of detection and not much bang for the buck.

PATHOPHYSIOLOGY
• The dissection occurs when there is a weakening of the tri-laminar aortic wall such that the intima is disrupted and the flow of high pressure blood then strips that layer from the media layer.
• Symptoms vary according to where the dissection occurs and progresses to.
• There are two basic classifications of dissections – Stanford and DeBakey – but the Stanford classification seems to be the best accepted.

CLASSIFICATION OF AORTIC DISSECTION
• There are two types of dissections in the Stanford classification:
  • Type A – involves the ascending aorta and/or arch, is the most common type, has the higher mortality, the mortality can be improved with surgery.
  • Type B – involves the arch distal to the take off of the left subclavian artery and/or the descending aorta and is not amenable to surgical repair.
• The American Heart Association classifies Type A as any dissection that involves the ascending aorta regardless of site of origin.
AORTIC ANATOMY

The ascending aorta has 3 main branches and note the origins of the coronary arteries.

As the aorta passes below the diaphragm numerous branches take off including those to the spinal cord, kidneys, and intestines.

PRESENTATION OF ATAD

• The classic presentation of ATAD is:
  - Sudden onset – 84% described a sudden onset of pain in the International Registry of Acute Aortic Dissection (IRAD) in > 2,000 cases.
  - Severe chest pain often described as a "tearing" sensation.
  - Radiation of the pain to the back.
  - Syncope is not uncommon but a highly non-specific symptom unless accompanied by some of the above symptoms.

PRESENTATION OF ATAD - IRAD STUDY

• This was a study of 464 patients, 62% with Type A, 38% Type B dissections.
• Severe, acute onset of pain was the most common symptom.
• Exam: aortic regurgitation 32%, pulse deficit only 15%.
• CXR normal 12%, EKG normal 31%, so often not helpful.
• Mortality: Overall 27%, Type A surgically managed 26%, Type A without surgery 58%, Type B medical management 11%, Type B surgery 31%.

PRESENTATION VARIABLES

• Depending on the location of the dissection and the other arteries it might involve symptoms can vary markedly.
  - Type B dissections often involve abdominal or back pain as mesenteric or spinal arteries are obstructed by the dissection.
  - Hypertension is seen more in type A dissections and may account for its increased mortality.
  - Chest pain and acute MI can be seen if the coronary artery ostia are occluded.
  - Most, if not all symptoms, of AAD are non-specific and thus can point to a number of different entities that have similar symptom patterns.

PRESENTATION VARIABLE

• Other clinical scenarios where AAD might be suspected:
  - Chest pain associated with pain below the diaphragm.
  - Chest pain associated with a neurologic deficit, typically in the legs as the dissection cuts off the spinal arteries.
  - Severe chest pain associated with acute myocardial infarction as the dissection cuts off the ostia of the coronary arteries just above the aortic valve.
  - Chest/abdominal pain in a high risk patient – to be discussed later.
ARE THERE SILENT PRESENTATIONS OF ATAD?

- Unfortunately there are significant number of patients with no symptoms of dissection in a study from Hong Kong of 141 patients with ATAD:
  - 43% had no chest pain with a trend toward "non-urgent" triage.
  - Thoracic aneurysm was more frequent (26% vs 13%) and more likely to present with abdominal pain or back pain.
  - Less likely to present with sudden onset of pain (31% vs 48%) but more likely to be in shock (21% vs 8%).
  - Less likely to present with ischemia on EKG (15% vs 36%) and less likely to get either an EKG or CXR.
  - Trend towards Type B dissections (38% vs 26%) with a higher 30 day mortality (15% vs 11%).


RISK ASSESSMENT

- As for ANY clinical entity for which one is going to test and work-up some idea of risk has to be a key first question.
  - With some 8-10% of all patients presenting to an emergency department with some type of chest pain very few will actually have an ATAD.
  - So, among a number of questions regarding non-traumatic acute aortic dissection addressed in an article by the ACEP Clinical Policies Subcommittee* on Aortic Dissection is there a clinical decision rule that can identify patient at low risk?

D-DIMER

- While a positive d-dimer was seen in 91-100% of ATAD patients one must keep in mind when that the test can be both false positive or negative.
  - False negatives (low d-dimer with ATAD) can be seen with a longer time from symptom onset, intramural thrombi, short length of dissection, or younger age.
  - A false positive (high d-dimer but no ATAD) can be seen with symptom mimics such as PE, AMI, older patients, or inflammatory conditions.
  - So, by itself d-dimer cannot reliably rule in or rule out a dissection.

WHAT ABOUT IF WE ALL PLAY TOGETHER?

- But, what if one combines a set of symptoms, physical exam findings, and some laboratory testing.
  - Can one then reliably risk stratify patients with ATAD in the differential diagnosis?
  - So, is there a risk stratification tool coupled with laboratory testing that can identify patients at risk for a dissection in whom imaging and/or transfer would be indicated?
AORTIC DISSECTION DETECTION RULE PLUS D-DIMER STUDY – ADVISED STUDY

• The AdviSED study was an international, multicenter, prospective study of 1,850 patients who presented within 14 days with >1 of: non-traumatic chest pain, abdominal pain, back pain, syncope, or perfusion deficit (CNS, mesenteric, myocardial, limb) (schematized) where ATAD was in the differential.

• Patients first underwent an Acute Aortic Dissection (AAD) Score and had a d-dimer level run.

• In those with an AAD Score <1 PLUS a negative d-dimer only 3 patients were found to have a ATAD – sensitivity 99.6%, NPV 99.7%.


ACUTE AORTIC DISSECTION (AAD) RISK SCORE

• AAD risk score looked at 3 areas: high risk conditions, high risk pain features, and high risk exam features.

  • High risk conditions: Marfans, FH of aortic disease, known AV disease, recent AV manipulation, known thoracic aneurysm.

  • High risk pain features: chest/back/abdominal pain of abrupt onset, severe, ripping or tearing quality.

  • High risk exam features: evidence of perfusion deficit (pulse deficit, SBP difference, focal neurologic deficit along with the pain), new aortic insufficiency murmur, hypotension or shock.

  • A point is given for any positive with a category so a maximum of 3 points.


ATAD + D-DIMER

• Using the data from the AdviSED study one could then risk stratify patients where ATAD is in the differential diagnosis as follows:

  • AAD score <1 with a negative d-dimer – look for another diagnosis besides ATAD.

  • AAD score <1 BUT positive d-dimer – consider work-up for ATAD.

  • AAD score >1 and positive d-dimer – needs to be worked-up for ATAD.

• Limitations of the study were the ED physician selected the patients to be included but a 13% incidence of ATAD suggests they were very careful in their selections along with an ultimate dependence on the d-dimer as the discriminating test.

• From the data in this study it would be prudent for a patient with an elevated AAD Score and elevated d-dimer (you might even skip the d-dimer with a high AAD score) or an AAD Score >1 but a positive d-dimer to either be transferred to a dissection capable institution or to image the patient and notify the cardiovascular surgeons of the potential of an ATAD.

IS THE ADVISED STUDY READY FROM PRIME TIME?

• Keep in mind the AdviSED Study had some limitations including:

  • The protocol has not undergone a verification study.

  • Had a very high incidence of ATAD – 13%, far higher than most other populations.

  • Not sure the study has as yet widespread applicability.

  • Likelihood ratios* for combining the AAD Score and D-dimer –

    • LR neg = 0.02

    • LR pos = 1.22

  • So a better LR neg (which is what you want to screen patients) but still not great.

*Likelihood ratios calculated by Raymond Jackson, MD; Wm. Beaumont Department of Emergency Medicine

WORK-UP FOR ATAD

• Imaging is the key element to diagnose a dissection.

  • The options are CTA, TEE, or MRA – clearly the availability of CTA far exceeds that of TEE or MRA but is it as accurate?

  • ACEP Clinical Policy was able to find a Level 2 (moderate clinical certainty) that CTA has an accuracy similar to that of TEE and MRA in particularly if one asks the reverse question – can a negative CTA exclude the diagnosis?

  • In a good Class 1 meta-analysis the sensitivity/specificity for CTA was 100%/98%, for TEE 98%/95%, and MRA 98%/98%.

  • In addition both CTA and MRI have the ability to detected alternative findings in patients without aortic disorders that TEE does not.
MRI Images of Type A and Type B dissections.

TEE IN ATAD

- TEE is becoming more available in EDs as emergency physicians advance their ability with ultrasound and some ED physicians are performing this test.
- The advantage is it can be done at the bedside and relatively rapidly through in some patients prior intubation might be required.
- The ACEP Clinical Policy determined a Level B recommendation suggested NOT to rely on abnormal bedside TEE to definitively establish the diagnosis but also had a Level C recommendation that with an abnormal TEE that surgical consultation and/or transfer of the patient be considered.

TEE Images of dissections

THERAPY FOR ATAD

- Therapeutic treatment of an ATAD depends on type.
- Type A has the potential for surgery so notification of cardiovascular surgery and/or transfer to a center capable of such surgery is extremely important.
- Type B dissections are treated medically so the urgency for surgical intervention is less.
- Keep in mind for Type A dissections mortality increases ~8% per hour so time is not on the patient's side - nor on yours.

TREATMENT IN THE ED

- Death from a dissection is typically due to rupture that becomes more likely as the intimal tear progresses.
- That progression is attributed to the kinetic energy resulting from the pulsatile nature of blood flow in the aorta along with the pressure behind that flow.
- Based on a study by Wheat in 1965 medications to reduce the heart rate and lower the blood pressure have been bedrock goals in the therapy for an ATAD and in Type A dissections to buy time until the patient can be taken to the OR.

Wheat suggested a BP goal of 100-120mmHg, but does a targeted heart rate and blood pressure reduce the mortality or morbidity of ATAD?

- Again the ACEP Clinical Guidelines attempted to address this key issue and were only able to find a Class C recommendation (expert opinion level) to decrease pulse and/or pressure if elevated in ATAD patients.

- However, there are no specific targets that have demonstrated a reduction in mortality or morbidity.

TREATMENT IN THE ED
BP AND PULSE GOALS

- The major specialty consensus guidelines suggest a heart rate goal of 60/minute with a blood pressure goal below 120mmHg, but limited data support this recommendation.
- In a Class III study of 171 patients followed for 2 years adverse events were lower in those who met BP and HR goals – but this was not a great study.
- Another study found a pressure above 140mmHg was not associated with an increase in aorta size – again not a great study.

MEDICATIONS

- Lowering both BP and pulse can be at odds with each other.
- As BP is lowered often there is a reflex increase in heart rate defeating the goal of reducing the shear force of the blood that is separating the intima and causing the dissection.
- As such often two medications are necessary – one to reduce heart rate and the other to lower the blood pressure.

MEDICATIONS

- Various recommendations of drug combinations have been suggested and the literature has not established the best medications to use but a beta-blocker combined with a blood pressure lowering agent are most often suggested.
- So here is a CLASS III recommendation from the presenter:
  - Beta-blocker – esmolol
  - Blood pressure lowering agent – nitroprusside
- Esmolol reduces the pressure while esmolol blocks the reflex increase in heart rate, blood velocity is reduced with a concomitant reduction in shear force.

ESMOLOL

- Esmolol is a beta-blocking agent whose advantages are:
  - Rapid onset – when a bolus is combined with an infusion within minutes.
  - Short duration of action – metabolized but enzyme that reside on the RBC’s, does not depend on hepatic or renal pathways for degradation or excretion.
  - The use of a bolus dose to saturate the RBC enzymes shortens the time to steady state substantially.
- Dose: bolus = 500 mcg/kg (0.5 mg/kg) over 1 minute, infusion starts at 50 mcg/kg/min, if goal not met increase infusion to 100, 150, 200 (max) mcg/kg/min – repeat the bolus dose between increases in infusion rate.

NITROPRUSSIDE

- In use in medicine since 1928, arterial and venous vasodilator (works on arterioles > venules as it is broken down to nitric oxide).
- It is metabolized to cyanide but in years of monitoring Beaumont has never seen a single incidence of toxicity and patients on infusions are no longer monitored for cyanide accumulation.
- Administered by infusion, can be given through a peripheral line.
- Dose: initial infusion rate = 0.3 mcg/kg/min titrating upwards to maximum of 10 mcg/kg/min.

OTHER POSSIBLE MEDICATIONS

- Labetalol – has both an alpha/beta blocking effect, in hypertension typically does not cause severe hypotension, onset = 2.5 minutes, half-life = 6-8 hours, so exactly titratable; dose = 20mg IV, repeat 40mg, then 80mg, maximum is 300mg total dose – used in ATAD not well established
- Nicardipine – a hypotensive agent, often used in hypertension and intracranial pathology, use in ATAD not established, typically does not bottom out the pressure, will not block increased pulse rate; dose = .5mg/hr to maximum of 1.5mg/hr.

*Presenters note: my own case N=1, patient failed nicardipine in terms of lowering the BP.*

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SURGERY FOR TYPE A DISSECTIONS

ATAD can start with an aneurysm, seen here, that is one of the risk factors with AAD Score.

SURGERY FOR TYPE A DISSECTIONS

Surgery most often consists of replacing the dissected aorta with a Gore-Tex graft.

SOMETIMES THINGS BLEED

This is high risk major surgery that can go sideways very rapidly.

SUMMARY

• Acute thoracic aortic dissection while an uncommon cause of chest pain has a high mortality and the time it takes to diagnose it can be without delays increase the mortality (~8%/hour in Type A dissections).
• Anatomically ATAD results when there is a tear in the intima and the pulsatile blood flow strips away the intima off the media layer of the aorta.
• Symptoms vary according to the level of the dissection and what arteries off the aorta are involved.

SUMMARY

• There are 2 types with the Stanford classification – Type A involving the ascending aorta, Type B involving the descending aorta beyond the take off the left subclavian branch.
• Type A dissections can be amenable to surgery so timely notification of cardiovascular surgery with transfer to a hospital capable of the surgery is paramount.
• Type B are treated medically primarily focusing on blood pressure control.

SUMMARY

• Symptoms – depends on where the dissection occurs:
  • Classically a sudden onset of severe chest pain, radiating to the back, the pain is severe and constant
  • Syncope, pain in both the back and below the diaphragm can be seen.
  • Chest pain associated with neurologic symptoms such as arm/leg paralysis.
  • Paroxysms of chest pain can cause chest and/or abdominal pain.
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• Perfusion deficits with pulse/BP inequality between the arms or legs is highly suggestive of ATAD.
SUMMARY

- Physical, X-ray, EKG, Lab Findings:
  - On exam a pulse deficit and/or blood pressure deficit > 20mmHg – seen occasionally but is highly correlated with ATAD.
  - CXR may show widening of the mediastinum – also strongly correlated with ATAD.
  - EKG is most often normal but with involvement of the coronary arteries changes with acute myocardial infarction can be seen.
  - Labs are most often non-specific or reflect the arterial branches of the aorta involved i.e. elevated troponin or lactate if the coronary or intestinal branches are occluded by the dissection.
  - D-dimer is often elevated in ATAD and can help risk stratify.

SUMMARY

- Acute Aortic Dissection Rule*: summarizes the potential risks:
  - High risk conditions: Marfans, FH of aortic disease, known AV disease, recent AV manipulation, known thoracic aneurysm.
  - High risk pain features: chest/back/abdominal pain of abrupt onset, severe, ripping or tearing quality.
  - High risk exam features: evidence of perfusion deficits (pulse deficit, SBP difference, focal neurologic deficit along with the pain), new aortic insufficiency murmur, hypotension or shock.

*1 point for any positive in a category – maximum of 3

SUMMARY

- The AAD Rule plus a d-dimer in the AdviSED Trial suggested a means of risk stratification and work-up:
  - AAD < 1 and d-dimer < 500 – ATAD unlikely and work-up not indicated.
  - AAD > 1 or d-dimer > 500 – ATAD should be strongly in the differential, consider work-up.
  - AAD > 1 and d-dimer > 500 – ATAD a primary consideration, start work-up.

SUMMARY

- Imaging work-up is the most definitive:
  - CT scan for dissection often is the most readily available study – highly accurate both positive and negative.
  - MRI is also an excellent test in demonstrating a dissection.
  - TEE while not as good as CT or MRI, still if positive would be sufficient to alert CV surgery, start transfer if necessary, and proceed to CT or MRI.

SUMMARY

- Treatment in the ED:
  - Reduce shearing force of the blood flow by:
    - Lower the blood pressure – nitroprusside.
    - Reduce the heart rate that often elevates as BP is lowered – esmolol
  - For Type A dissections rapid notification of CV surgery and/or transfer to a hospital capable of providing the surgery.
  - For Type B dissections medical management with aggressively treating the blood pressure is the mainstay of treatment.

Well, hopefully I have not put you to sleep like I apparently have Winston.

Thank you.

Contact me with any questions: blwalters@beaumont.edu