Merito di una puttana
The tragedies of life are largely arterial.

(Osler)
Acute Aortic Syndrome

Classic dissection

Intramural haematoma → Aortic ulcer

VILACOSTA, I. et al. Heart 2001;85:365-368
Classic AD, with true and false lumen

Intramural haematoma

Subtle or discrete with bulging of the aortic wall

Ulceration of aortic plaque

Jatrogenic
Un’ intima debolezza
<table>
<thead>
<tr>
<th>De Bakey</th>
<th>Type I</th>
<th>Type II</th>
<th>Type III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stanford</td>
<td>Type A</td>
<td>Type A</td>
<td>Type B</td>
</tr>
</tbody>
</table>

Ascending dissections are almost twice the number of cardiac surgical emergencies.
The **DISSECT** mnemonic communicates fundamental anatomical and clinical features to select between medical management, open surgical repair, or endovascular interventions.

The classification has six elements:

- **duration of dissection (D)** less than 2 weeks, 2 weeks to 3 months, and more than 3 months from initial symptoms;
- **intimal tear location (I)** in the ascending aorta, arch, descending, abdominal, or unknown;
- **size of the aorta (S)**;
- **segmental extent (SE)**;
- **clinical complications (C)** such as aortic valve compromise, tamponade, rupture, and branch malperfusion; and
- **thrombosis (T)** of the false lumen and the extent.
Penetrating aortic ulcer

older age, male gender, tobacco smoking, hypertension, CAD, COPD, and concurrent abdominal aneurysm.

Symptoms have to be assumed to indicate an emergency as the adventitia is reached and aortic rupture expected.

Propagation of the ulcerative process may either lead to IMH, pseudoaneurysm, or even aortic rupture, or an acute AD.

European Heart Journal (2014) 35, 2873–2926
Intramural haematoma

Aortic IMH is an entity within the spectrum of AAS, in which a *haematoma develops in the media* of the aortic wall in the *absence of an FL and intimal tear*.

The *mortality rates of medically treated* patients in European and American series are high.

European Heart Journal (2014) 35, 2873–2926
Rara è rara.

E questo è un (il) problema.
The EP must walk a careful line between the significant risks of missing the diagnosis and the considerable clinical and financial burden of overtesting for this rare entity.
3 cases per 100,000 per year, which is half the incidence of symptomatic aortic aneurysm.

Su incidencia es de unos 30 casos por millón de habitantes al año, de los cuales el 80% son disecciones, el 15% hematomas intramurales y el 5% úlceras penetrantes.

Evangelsista A. Tratado de Cardiologia, 2015
Given the **low number** of patients who present to the ED with a thoracic aortic dissection as well as the **variability in presentation**, a prospective analysis of the aortic dissection detection risk score would be challenging.

It is possible that the **specificity** could be very low, possibly to the point of being **prohibitive**.

- **440** per 100,000 for **ACS**
- **69** per 100,000 for **PE**
- **3 to 4** per 100,000 for acute **AD**

Lancet 2015; 385: 800–11
**Availability:** the disposition to judge things as being more likely, or frequently occurring, if they readily come to mind. Thus, *recent experience with a disease may inflate the likelihood of its being diagnosed.*

**Base-rate neglect:** the tendency to ignore the true prevalence of a disease, either inflating or reducing its base-rate, and distorting Bayesian reasoning. However, in some cases, clinicians may (consciously or otherwise) *deliberately inflate the likelihood of disease,* such as in the strategy of “rule out worst-case scenario” to avoid missing a rare but significant diagnosis.

Sempre in cerca di un pretest, in un mondo senza regole

There are no prospectively tested rules to risk stratify chest pain for the risk of AD.

Class I
Providers should routinely evaluate any patient presenting with complaints that may represent acute thoracic AD to **establish a pretest risk of disease** that can then be used to guide diagnostic decisions

AHA GL, Circulation. 2010;121:1544-1579
Likelihood ratio

\[
LR^+ = \frac{Sens}{1 - Spec}
\]

\[
LR^- = \frac{1 - Sens}{Spec}
\]
Risk Factors for Development of Thoracic AD

**Conditions associated with increased aortic wall stress**
- Hypertension, particularly if uncontrolled
- Pheochromocytoma
- Cocaine or other stimulant use (...energy drinks...)
- Weight lifting or other Valsalva maneuver
- Trauma: Deceleration or torsional injury (eg, motor vehicle crash, fall)
- Coarctation of the aorta

**Conditions associated with aortic media abnormalities**

*Genetic*
- Marfan, Ehlers-Danlos, Loeys-Dietz vascular form
- Bicuspid aortic valve (including prior aortic valve replacement) Turner syndrome
- Familial thoracic aortic aneurysm and dissection syndrome

*Inflammatory*
- Vasculitides Takayasu, Giant cell arteritis, Behçet arteritis

*Other*
- Pregnancy
- Polycystic kidney disease
- Chronic corticosteroid or immunosuppression agent administration
- Infections involving the aortic wall either from bacteremia or extension of adjacent infection

*Circulation.* 2010;121:1544-1579
Clinically AD will often present as a **two step process**.

After a **first event** which is associated with severe pain and pulse loss, **the bleeding stops**. The **second event** sets in when the pressure exceeds a critical limit and **rupture occurs**, either into the pericardium with cardiac tamponade or into the pleural space or mediastinum.

Task Force on Aortic Dissection, European Society of Cardiology
Propagation of the dissection can occur both distal and proximal to the initial tear, involving branch vessels and the aortic valve and entering the pericardial space. Such propagation is responsible for many of the associated clinical manifestations.
SINTOMATOLOGIA

• Classica

Dolore (95%) ben localizzato (64%), tipo pugnalata, strappo (51%) ad insorgenza improvvisa (85%) al massimo della intensità fin dall’inizio, in sede toracica anteriore (73%), posteriore (36%) o addominale (30%), associato +/- a sintomatologia neurologica con +/- alterazioni dell’esame obiettivo.

• Atipica

Sincope senza dolore o deficit neurologici, Paraplegia, ,Insufficienza renale acuta, addome acuto, TIA, ictus,

Vomito, crisi convulsiva, diarrea, febbre
La gran puttana e
Le sindromi ischemiche associate:

- Coronarica
- Cerebrale
- Splancnica
- Arti
- ......
### Negative findings (patient doesn’t have)

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Decreased Disease Probability (Negative Likelihood Ratio)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sudden Chest Pain</td>
<td>$0.3 \times (0.2-0.4)$</td>
</tr>
<tr>
<td>History of Hypertension</td>
<td>$0.4 \times (0.3-0.6)$</td>
</tr>
<tr>
<td>Tearing/Ripping Pain</td>
<td>$0.4 \times (0.3-0.5)$</td>
</tr>
<tr>
<td>Migrating Pain</td>
<td>$0.6 \times (0.5-0.7)$</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Signs on Physical Exam</th>
<th>Decreased Disease Probability (Negative Likelihood Ratio)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulse deficit</td>
<td>$0.63 \times (0.4-1.0)$</td>
</tr>
<tr>
<td>Focal Neuro deficit</td>
<td>$0.87 \times (0.8-0.9)$</td>
</tr>
<tr>
<td>Diastolic murmur</td>
<td>$1.1 \times (0.6-1.7)$</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Lab and Study Findings</th>
<th>Decreased Disease Probability (Negative Likelihood Ratio)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enlarge aorta or wide mediastinum</td>
<td>$0.13 \times (0.02-1.00)^*$</td>
</tr>
<tr>
<td>LVH on admission EKG</td>
<td>$0.8 \times (0.2-0.9)$</td>
</tr>
</tbody>
</table>

### Positive findings (patient has this)

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Increased Disease Probability (Positive Likelihood Ratio)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tearing/Ripping Pain</td>
<td>$10.8 \times (5.2-22.0)$</td>
</tr>
<tr>
<td>Migrating pain</td>
<td>$7.6 \times (3.9-18.0)$</td>
</tr>
<tr>
<td>Sudden Chest Pain</td>
<td>$2.6 \times (2.0-3.5)$</td>
</tr>
<tr>
<td>Hx of Hypertension</td>
<td>$1.5 \times (0.8-3.0)$</td>
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</tbody>
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<tr>
<th>Signs on Physical Exam</th>
<th>Increased Disease Probability (Positive Likelihood Ratio)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Focal Neurologic Deficit</td>
<td>$33.0 \times (2.0-549.0)$</td>
</tr>
<tr>
<td>Diastolic Heart Murmur</td>
<td>$4.9 \times (0.6-40.0)$</td>
</tr>
<tr>
<td>Pulse Deficit</td>
<td>$2.7 \times (0.7-9.8)$</td>
</tr>
</tbody>
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<tr>
<th>Lab and Study Findings</th>
<th>Increased Disease Probability (Positive Likelihood Ratio)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enlarge Aorta or Wide Mediastinum</td>
<td>$3.4 \times (2.4-4.8)$</td>
</tr>
<tr>
<td>LVH on Admission EKG</td>
<td>$0.8 \times (1.5-4.0)$</td>
</tr>
</tbody>
</table>
• The absence of pain of sudden onset substantively decreases the probability of dissection (negative LR 0.3; 95% CI, 0.2-0.5)

• tearing or ripping pain (positive LR 1.2-10.8)

• pain that migrates (positive LR 1.1-7.6).

Does This Patient Have an Acute Thoracic Aortic Dissection?

*JAMA.* 2002;287:2262-2272.
Come l’ESA.

Un dolore mai provato prima....
About 10% of aortic dissections are **painless** and may present with symptoms secondary to complications of the dissection.

Emerg Med J 2006;23:e24
1 malessere generale epigastralgia shock tamponamento
2 **dolore toracico**
3 *sincope*
4 dolore addominale deficit arti inferiori
5 **dolore toracico**
6 **dolore toracico** dispnea
dolore toracico e addominale shock
dolore toracico e addominale
dolore toracico *sincope*
dolore toracico
7 *sincope*
8 **dolore toracico** *sincope*
9 shock
10 **dolore toracico**
11 *sincope*
12 **dolore toracico** *sincope*
13 shock

13 accessi consecutivi per DA, PS Udine
A better history may increase the diagnostic yield.

Any patient with a combination of 

**cardiac and neurological symptoms**

should be considered at high risk for aortic dissection.
A recent history of strenuous exercise or use of drugs (such as cocaine or amphetamines) followed by severe chest back pain is highly suggestive of acute aortic dissection.

Lancet 2015; 385: 800–11
Physical findings associated with TAD tend to be present in a third or fewer cases; however, pulse deficits (positive LR, 5.7; 95% CI, 1.4-23.0) or focal neurological deficits (positive LR, 6.6-33.0) greatly increase the likelihood of thoracic AD in the appropriate clinical setting.

The presence or absence of a diastolic murmur is not useful (positive LR, 1.4; negative LR, 0.9).

Does This Patient Have an Acute Thoracic Aortic Dissection?

*JAMA.* 2002;287:2262-2272.
Hypertension is a typical finding.

Patients with normal or low BP should be assumed to be in shock until proven otherwise.

Hypotension is more commonly associated with a type A dissection and is also associated with a high rate of mortality in the acute setting.

Amal Mattu & C
Evoiding Common Errors in ED

Overall the clinical examination is insufficiently sensitive to rule out AD given the high morbidity of missed diagnosis.

Does This Patient Have an Acute Thoracic Aortic Dissection?  
*JAMA.* 2002;287:2262-2272.
ECG

The ECG changes in AD are usually nonspecific, but 1% to 2% of patients have acute ST elevation.

*Circulation.* 2010;122:184-188
RX TORACE

- Possibili alterazioni (60-90%)
  - Allargamento bottone aortico o mediastino
    - (63% DA tipo A e 56% DA tipo B)
  - Spostamento calcificazioni intimali
  - Versamento pleurico (sn>dx)
  - Opacizzazione “finestra AP” (spazio tra aorta e arteria polmonare)
  - Addensamento pleurico apicale sinistro (CAP)
  - Contorno irregolare o indistinto dell’aorta
  - Deviazione esofagea o tracheale
A normal aorta and mediastinum on chest radiograph helps to exclude the diagnosis (negative LR, 0.3; 95% CI, 0.2-0.4).
Utility of CXR is to help further lower your pretest probability in the low risk patient.

...a completely normal chest radiograph result or the absence of pain of sudden onset lowers the likelihood.

Does This Patient Have an Acute Thoracic Aortic Dissection?
Fatelo in piedi
DD has a sensitivity of 97% and a specificity of 47%, implying that a negative DD test can probably exclude the diagnosis of acute dissection.

D-dimer analysis has sufficient predictive value to guide the need for further imaging.

Lancet 2015; 385: 800–11
Because DD is nonspecific, routinely obtaining this test in a large population of patients with symptoms suspicious for AD can result in harm, most notably, exposure to radiation and cost associated with advanced imaging.
The following conditions may result in a low or false-negative DD value in patients with proven thoracic AD:

- chronicity,
- time from symptom onset,
- presence of thrombosis or intramural hematoma,
- short length of dissection, and
- young age of patient.
For **non-low-risk patients** D-dimer testing was not sufficiently sensitive to rule out AD and **should not be used**.

D-dimer levels are best used for ruling out AAD in patients with low likelihood of the disease.

*do not rely* on DD*alone* to exclude the diagnosis of aortic dissection.

Medicine 2015; 94(4):e471

• muscolo liscio vascolare (catene pesanti della miosina),

• interstizio vascolare (calponina),

• lame elastiche (frammenti solubili dell’ elastina)
In adult patients with suspected nontraumatic thoracic aortic dissection, EP may use CTA to exclude thoracic aortic dissection because it has accuracy similar to that of TEE and MRA.

TOE may be of great interest in the very unstable patient, and can be used to monitor changes in-theatre and in post-operative intensive care.
La mejor combinación para un correcto diagnóstico de la disección aórtica aguda y sus complicaciones es la realización de una TC y una ETT.

Evangelsista A. Tratado de Cardiología, 2015
Remember to aggressively manage BP in patients with acute thoracic AD

Amal Mattu & C
Evoiding Common Errors in ED
...decrease blood pressure and pulse if elevated.

However, there are no specific targets that have demonstrated a reduction in morbidity and mortality.
Progression of dissection has been attributed to the pulsatile nature of blood flow.

…the pulsatile flow is a result of 2 key forces that may be targeted:

- the **kinetic energy** of the blood flow that can be reduced if the velocity of the blood flow is reduced
- the **pressure differentials** throughout the aorta.

Wheat MW Jr, Palmer RF, Bartley TD, et al.


Wheat MW Jr, Palmer RF.

Prog Cardiovasc Dis. 1968;11:198-210
Rara e amara

Cambiare mestiere?