Constriction/ Restriction

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Differentiation of Constriction vs. Restriction

• Why is it important?

• Important therapeutic implications
  • Pericardiectomy
  • Heart failure management and treatment of underlying disease
Outline

• Normal pericardium

• Restriction cardiomyopathy

• Constrictive pericarditis

• Differentiating between the two processes
Pericardium

- Fibroelastic sac surrounding the heart
- Two layers
- Physiologic pericardial fluid <50 mL
Normal Pericardium

Inspiration

Intrathoracic pressure decreases

Increase in venous return to the right heart

Transient increase in RV size

(No impairment of LV filling)
Normal Pericardium

- Expire
- Intrathoracic pressure increases
- Increase in pulmonary venous return to the left atrium
- LV filling
Restrictive Cardiomyopathy

• Disease of the myocardium

• Predominant diastolic, rather than systolic, dysfunction

• Pulmonary systolic pressure usually is moderately to severely elevated
Clinical Manifestations of Restrictive Cardiomyopathies

- Exercise intolerance
  - Impaired ability to augment cardiac output with tachycardia because diastolic restriction of filling

- Peripheral edema, hepatomegaly, ascites, anasarca

- Highly prone to developing atrial fibrillation
Restrictive Cardiomyopathies

Noninfiltrative
- Idiopathic
- Hypertrophic
- Radiation
- Eosinophilic

Infiltrative
- Amyloidosis
- Sarcoidosis
- Gaucher disease
- Hurler disease

Storage Disease
- Hemochromatosis
- Fabry Disease
- Glycogen storage disease
Typical Echocardiographic Features

- Non-dilated thick-walled LV
- Abnormal diastolic function
- RV free wall thickening
- Bi-atrial enlargement
- Elevation of pulmonary pressures
- Elevation of RA pressures
Doppler Features of Restrictive Cardiomyopathy

- Mitral inflow E/A ratio > 2.5
- DT of E velocity < 150 msec,
- IVRT < 50 msec
- Decreased septal and lateral e’ velocities (3–4 cm/sec)
  - Lateral > septal e’ velocity
- E/e’ ratio > 14
- Inc LA volume index > 50 mL/m²
E/A = 1.7
DT = 140
E’ = 4
E/E’ = 17
Amyloid Heart Disease
Amyloid Heart Disease

• Disorder of protein metabolism which results in protein deposition in organs and tissues

• Amyloid deposition begins in sub-endocardium and extends within the myocardium between the muscle fibers
# Main Types of Amyloidosis

## Isolated Deposits

<table>
<thead>
<tr>
<th>Type</th>
<th>Source of Amyloid</th>
<th>Organs Involved</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>AL (Primary)</strong>&lt;br&gt;Amyloidosis&lt;br&gt;Amyloid Light-Chain</td>
<td>Bone Marrow&lt;br&gt;(Light chains produced by plasma cells)</td>
<td>Kidneys, Heart, Liver, GI system, Nervous system</td>
</tr>
<tr>
<td><strong>AA (Secondary)</strong>&lt;br&gt;Amyloidosis&lt;br&gt;Amyloid A Protein</td>
<td>Circulating inflammatory protein&lt;br&gt;(Serum amyloid A)</td>
<td>Kidneys, Liver</td>
</tr>
<tr>
<td><strong>TTR (Familial)</strong>&lt;br&gt;Amyloidosis&lt;br&gt;Mutant Transthyretin</td>
<td>Unstable, mutant transthyretin&lt;br&gt;produced in the liver</td>
<td>Nervous system, Heart</td>
</tr>
<tr>
<td><strong>SSA (Senile systemic)</strong>&lt;br&gt;Amyloidosis&lt;br&gt;Seniors</td>
<td>Wild-type (normal) transthyretin</td>
<td>Heart</td>
</tr>
</tbody>
</table>
Echocardiographic Features of Amyloid Infiltration

- Increased LV wall thickness ("ground glass")
- Increased RV wall thickness

- Small LV; normal or reduced systolic function
  - Pericardial effusion; big LA; thick atrial septum
  - Valve thickening; mild regurgitation

- E/A ratio >2
  - Decleration time <150 ms
  - PV: small S wave, large D wave; S/D ratio <0.5
Case 1

76 year old man with senile cardiac amyloidosis, Afib, ICD presents with generalized weakness
Case 2

35 year old man being evaluated for heart/liver transplant for history of hemochromatosis
Hemochromatosis

- Initially characterized by diastolic dysfunction and arrhythmias and in later stages by dilated cardiomyopathy

- Diagnosis of iron overload is established by elevated transferrin saturation (>55%) and elevated serum ferritin (>300 ng/mL)

- Genetic testing for mutations in the HFE gene
Hemochromatosis
CMR T2-Weighted Images. Both the myocardium & liver (L) show decreased signal intensity compared to trapezius (T) skeletal muscle.
Case 3

37 year old man with hypertension, end stage renal disease on dialysis, and Fabry’s disease on RV biopsy
Fabry’s Disease

- X-linked, lysosomal storage disease
- Dysfunctional metabolism of sphingolipids
- Mutation in GLA gene
  - Makes enzyme α-galactosidase A
  - Build up of globotriaosylceramide

- Symptoms/clinical findings
  - Acroparesthesias
  - Angiokeratomas
  - Hypohidrosis
  - Corneal opacity
  - Tinnitus and hearing loss
  - Progressive kidney damage
  - Cardiomyopathy
  - Stroke
Constrictive Pericarditis
Constrictive Pericarditis

• Long-standing inflammation that leads to pericardial scarring with thickening, fibrosis, calcification

• Loss of normal elasticity of the pericardial sac

• Characteristic hemodynamic changes occur from changes in intrathoracic respiratory pressure with a fixed end-diastolic ventricular volume
Constrictive Pericarditis

- **Etiology**
  - Idiopathic
  - Post-infectious: TB, viral, other
    - Recurrent pericarditis
  - Cardiac surgery
  - Radiation
  - Trauma
  - Malignancy

- **Signs**
  - Right >> Left HF
  - Elevated JVP with prominent Y descent
  - Kussmaul’s sign
  - Pericardial knock
  - Edema
  - Ascites
Calcified Pericardium
Figure 5. Chest CT from a patient with pericardial constriction showing thickened pericardium (arrows) and a left pleural effusion.
Constrictive Pericarditis

Inspiration

Expiration
Constrictive Pericarditis: Pathophysiology

• Marked restriction of filling
• Elevation and equilibration of filling pressures in all heart chambers
  • Early to mid diastole ventricular filling is abrupt and rapid
  • This filling abruptly ceases when the intracardiac limit reaches its set limit
• Systemic venous congestion leads to hepatic congestion, peripheral edema, ascites, and sometimes anasarca
With tamponade, diastolic filling is impaired in both early and late diastole due to the elevated pericardial pressures “compressing” the heart.

With constriction, early diastolic filling is rapid but ends abruptly when the volume limits of the rigid pericardial space are reached.
Inspiration  Expiration

Braunwald’s Heart Disease.
Constriction: Hemodynamics

- RA: elevated pressure with “M” or “W” pattern
- RV: “dip and plateau” or “square root” sign
- Diastolic equalization of LV and RV pressures
  - Volume loading
- Systolic discordance of LV and RV pressures
Tamponade

Constriction
Constriction: Diastolic Equalization

“Dip and plateau”
Echo findings in Constriction

• LV size and systolic function typically normal

• M-mode may show persistent pericardial thickening with low-gain settings

• >25% inc in MV inflow velocities seen with expiration

• Septal bounce

• IVC and hepatic vein plethora
  – Diastolic HV flow reversal

• Mitral E wave velocity usually < 160 msec

• Typically normal PASP
M-mode

Thick pericardium that persists at low gain settings
Doppler Findings in Constriction

- Abnormal passive filling of the ventricles during early diastole $\rightarrow$ High E velocity
- Tissue Doppler: annulus reversus
- Respiratory variation in ventricular filling
  - Inspiration
    - MV inflow: decreases $\geq 25\%$
    - TV inflow: increases $\geq 40\%$
- Hepatic venous flow reversal with expiration
Normal

Constriction

Restriction

Annulus Reversus
Tricuspid Valve In-Flow

- Tricuspid inflow
  - E wave increase ≥ 40% with inspiration
Hepatic vein

- **Expiration:** Enhanced diastolic flow reversal
Diastolic Septal Bounce

Rapid filling during early diastole leads to asymmetrical filling of the RV and LV creating a fluctuating pressure gradient and an abrupt shift of the septum.
Dilation and lack of respiratory variation in IVC
Differentiation of Constriction vs. Restriction

- Different etiologies
## Differentiation of Constriction vs. Restriction

<table>
<thead>
<tr>
<th></th>
<th>Constrictive pericarditis</th>
<th>Restrictive cardiomyopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mechanisms of disease</strong></td>
<td>Cardiac volume constrained by inelastic pericardium resulting in impaired ventricular filling</td>
<td>Restriction of filling from impaired ventricular diastolic filling</td>
</tr>
<tr>
<td><strong>Physiological response</strong></td>
<td>Changes in intrathoracic pressure not transmitted to cardiac chambers (obliteration of pericardial space)</td>
<td>Normal respiratory variation in intrathoracic pressure transmitted normally to cardiac chambers</td>
</tr>
<tr>
<td><strong>Ventricular interaction</strong></td>
<td>Greatly enhanced</td>
<td>Unchanged</td>
</tr>
<tr>
<td><strong>Intrinsic myocardial function</strong></td>
<td>Normal</td>
<td>Abnormal</td>
</tr>
</tbody>
</table>
Differentiation of Constriction vs. Restriction

- Different etiologies
- Similar clinical presentations
- Similar physical exam signs
- Thick pericardium is not necessary or sufficient to dx constriction
- Overlapping echo & hemodynamic features
<table>
<thead>
<tr>
<th></th>
<th><strong>Constriction</strong></th>
<th><strong>Restriction</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Prominent y decent in venous pressure</td>
<td>Present</td>
<td>Variable</td>
</tr>
<tr>
<td>Paradoxical pulse</td>
<td>~1/3 cases</td>
<td>Absent</td>
</tr>
<tr>
<td>Pericardial knock</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Equal Right and Left sided pressures</td>
<td>Present</td>
<td>Left at least 3-5 mmHg &gt; right</td>
</tr>
<tr>
<td>Filling Pressures &gt; 25 mmHg</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>PASP &gt; 60 mmHg</td>
<td>No</td>
<td>Common</td>
</tr>
<tr>
<td>Hepatic veins</td>
<td>Inc expiratory flow reversal</td>
<td>Inc inspiratory flow reversal</td>
</tr>
<tr>
<td>“Square root” sign</td>
<td>Present</td>
<td>Variable</td>
</tr>
<tr>
<td>Respiratory variation inflows velocities</td>
<td>Exaggerated</td>
<td>Normal</td>
</tr>
<tr>
<td>Ventricular wall thickness</td>
<td>Normal</td>
<td>Usually Increased</td>
</tr>
<tr>
<td>Atrial Size</td>
<td>Possible LA enlargement</td>
<td>Bi-atrial enlargement</td>
</tr>
<tr>
<td>Septal Bounce</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Tissue Doppler e’ velocity</td>
<td>Increased</td>
<td>Reduced</td>
</tr>
<tr>
<td>Pericardial thickness</td>
<td>Increased</td>
<td>Normal</td>
</tr>
</tbody>
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Constriction
- Deformation of the LV and early diastolic recoil were attenuated in the circumferential direction

Restriction
- Attenuated in the longitudinal direction
# Differentiation of Constriction vs. Restriction

## Echocardiographic Parameters

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<tr>
<th>Parameter</th>
<th>Constrictive pericarditis</th>
<th>Restrictive cardiomyopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Septal bounce</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>MV inflow respiratory variation</td>
<td>≥25%</td>
<td>None</td>
</tr>
<tr>
<td>TV inflow respiratory variation</td>
<td>&gt;40%</td>
<td>None</td>
</tr>
<tr>
<td>MVDT</td>
<td>Short</td>
<td>&lt;160 ms</td>
</tr>
<tr>
<td>Hepatic vein reversal</td>
<td>Diastolic reversal with expiration</td>
<td>No change</td>
</tr>
<tr>
<td>IVRT</td>
<td>Decrease during expiration</td>
<td>No change</td>
</tr>
<tr>
<td>Increase during inspiration</td>
<td></td>
<td></td>
</tr>
<tr>
<td>TR duration</td>
<td>Increase</td>
<td>No change</td>
</tr>
<tr>
<td>E:e’</td>
<td>&lt;8–10</td>
<td>&gt;15</td>
</tr>
<tr>
<td>Myocardial mechanics with strain image</td>
<td>Normal longitudinal strain</td>
<td>Decrease longitudinal strain</td>
</tr>
<tr>
<td>Decrease net-twist angle</td>
<td>Normal net-twist angle</td>
<td></td>
</tr>
</tbody>
</table>

*E: Peak transmirtal flow velocity at early diastolic filling phase; e’: Peak early diastolic mitral annular velocity; E:e’: Ratio of E and e’ velocities; IVRT: Isovolumic relaxation time; MV: Mitral valve; MVDT: Mitral valve deceleration time; TR: Tricuspid regurgitation; TV: Tricuspid valve.*
Summary

• Restrictive cardiomyopathy is a disease of the myocardium and may be due to various etiologies including noninfiltrative, infiltrative, and storage diseases.

• Constrictive pericarditis is a disease of the pericardium leading to thickening and impairment of diastolic filling.
Summary

• There are numerous overlapping physical exam, echocardiographic and hemodynamic findings that overlap between the two disease states.
Summary

Think restrictive cardiomyopathy

- Thick walls, biaatrial enlargement
- Decleretion time <150 ms
- Decreased septal and lateral e’ velocities
  - Lateral E’ velocity > septal
- Elevated PASP
Think constrictive pericarditis

- Septal bounce
- Inspiratory drop in left heart velocities (MV, PV, LVOT)
- Thickened pericardium (not necessary)
- Dilated IVC
- Expiratory hepatic vein diastolic flow reversal
Thank you!