The RVSP is up: Now what?

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Disclaimer

- I have received honoraria from Actelion Pharmaceuticals Pfizer for speaking engagements as well as participation in advisory boards for GSK, Pfizer, Lilly, and Actelion.
- Assistance for participation in educational activities has also been received from Actelion and Pfizer.

PH referral Case

- 64 year old male
- Referred from a community respirologist in light of an echo reporting an elevated RVSP (56)
- Repeat echo at our institution:
  - RVSP 34
  - No evidence of RV dysfunction
  - WHO III
- Fibrotic NSIP
  - FVC 2.07 (57% predicted)

Q 1: Do you think that this person has pulmonary hypertension?

1) No
2) Yes mPAP 25 – 34 mmHg
3) Yes mPAP 34 - 40 mmHg
4) Yes mPAP 40+ mmHg
5) Yes but I have no idea what the mPAP is likely to be

Q 2: Would you send this patient for a right heart catheterization?

1) Yes
2) No

Q 3: If this patient was found to have PH by RHC would you offer targeted PH therapy?
(assume normal LVEDP)

1) No
2) Yes
3) Yes but only if the mPAP was >30
4) Yes but only if the mPAP was >35
5) Yes but only if the mPAP was >40
What to do with an elevated RVSP

- In the community (and PH centres) the primary screening tool for PH is echocardiography
- Most common reason for referral to the PH clinic:
  
  Elevated RVSP on echo

What does an elevated RVSP mean?

What does it not mean:

- RVSP ≠ mPAP
- Elevated RVSP ≠ PH
- PH is defined by the mPAP not the sPAP
- If the RVSP is to be used as an indicator of PH we must be able to predict the mPAP from the RVSP
- Does the echo derived RVSP correlate with the RHC sPAP?
- What is the relationship between the sPAP and the mPAP?

Correlation between echo derived RVSP and sPAP on RHC

Meta analysis of 27 studies

- Correlation coefficient 0.70 (0.67, 0.73)
- Sensitivity of 83% (73, 90)
- Specificity of 72% (53, 85)
- Positive likelihood ratio 3.0 (1.7, 5.3)
- Negative likelihood ratio 0.24 (0.14, 0.39)

Chemla Chest 2004
Chemla Chest 2009

Correlation between sPAP and mPAP

- mPAP = 0.61 x sPAP + 2 mmHg
- 25 = (0.61)x(37.7) + 2

Chemla Chest 2004
Chemla Chest 2009
Correlation sPAP and mPAP

\[ SPPAP = 1.50 \times mPAP + 2.46 \]

What to do with an elevated RVSP

- Have faith in the RVSP…
  - …but not unconditional trust
- Interpret the RVSP in context
  1) What evidence is there to support the diagnosis of PH?
  - How likely do you think it is that they have PH?
  2) What are their risk factors for PH?
  3) What information is missing?
  4) What is their functional class

Collateral information from echo

Common measurements

- Increased right heart chamber dimensions
- Increased RV wall thickness
- Abnormal shape and function of the interventricular septum
- Dilated main PA
- Short acceleration time of the RV ejection

2009 Dana Point Classification of PH

- Group 1: Pulmonary Arterial Hypertension
- Group 2: Left Heart Disease
- Group 3: Lung Disease and/or Hypoxia
- Group 4: Chronic Thromboembolic Pulmonary Hypertension
- Group 5: Unclear Multifactorial Mechanisms

Probability of PH based on echo and clinical information

<table>
<thead>
<tr>
<th>Low probability for PAH diagnosis</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Echocardiographic diagnosis of PH unlikely</td>
<td>no</td>
<td>I C</td>
</tr>
<tr>
<td>symptoms: no additional work-up is recommended</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Echocardiographic diagnosis of PH unlikely</td>
<td>presence of symptoms and/or associated conditions or risk factors for group 1—PAH: echocardiographic follow-up is recommended</td>
<td>I C</td>
</tr>
<tr>
<td>Echocardiographic diagnosis of PH unlikely</td>
<td>presence of symptoms, and absence of associated conditions or risk factors for group 1—PAH: evaluation of other causes for the symptoms is recommended</td>
<td>I C</td>
</tr>
</tbody>
</table>
Probability of PH based on echo and clinical information

Intermediate probability for PAH
- Echocardiographic diagnosis of PH possible
- No symptoms, and absence of associated conditions or risks factors for group 1—PAH: echocardiographic follow-up is recommended
- Echocardiographic diagnosis of PH possible
- Presence of symptoms, and absence of associated conditions or risks factors for group 1—PAH: RHC may be considered
- Echocardiographic diagnosis of PH possible
- Presence of symptoms, and absence of associated conditions or risks factors for group 1—PAH: alternative diagnosis and echocardiographic follow-up may be considered. If symptoms at least moderate RHC may be considered

High probability for PAH
- Echocardiographic diagnosis of PH likely
- With symptoms and presence/absence of associated conditions or risks factors for group 1—PAH: RHC is recommended
- Echocardiographic diagnosis of PH likely
- Without symptoms and presence/absence of associated conditions or risks factors for group 1—PAH: RHC should be considered

Should we consider RHC for patients if they will not be eligible for therapy?

Diagnostic Algorithm
- Non-invasive assessment compatible with PH
- Consider cause
- If group 2 or 3 AND "proportional" to severity
  - Treat underlying cause
- Look for Group 4 (V/Q)
- Perform RHC

What is “out of proportion” PH
- ECS - ERS - ISHLT (EHJ 2009)
- Group 2: TPG >12
- Group 3: Dyspnea insufficiently explained by lung mechanical disturbances and mPAP ≥ 40-45
- PVR or TPG is significantly elevated
- Dana Point Guidelines (JACC 2009)
- Group 2: TPG >12 and PVR > 3 WU
- Group 3: mPAP 35 - 50

Back to the case
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Back to the case

- May have PH
  - But if present is likely mild
- Most likely due to Interstitial Lung Disease (Group 3)
- No evidence of RV dysfunction
- Functional limitation in keeping with ILD severity
- Unless PH was out of proportion to ILD, PH therapy would not be offered
- Therefore: observe and follow with echos
- Do you agree or disagree?

Conclusion

- Elevated RVSP ≠ pulmonary hypertension
- Elevated RVSP ≠ need for right heart cath
- Elevated RVSP = look for PH
- RVSP must be interpreted in the context of collateral information
  - Risk of PH
  - Symptoms/history
  - Other echo findings
  - Other investigations