Lung Transplantation and PAH

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FOR IMMEDIATE RELEASE
Pioneer of Pulmonary Hypertension Treatment to be Honored at International PH Conference
SILVER SPRING, MD—On June 21, Robyn J. Barst, M.D., of New York, NY, will be recognized by the Pulmonary Hypertension Association (PHA) for her pioneering work in promoting clinical care, research, education and advocacy on behalf of patients with pulmonary hypertension (PH), a rare and deadly heart-lung disease. The PHA Award for Excellence in PAH Care will be presented to Dr. Barst at the Pulmonary Hypertension Association’s (PHA) 8th International PH Conference in Houston, TX.

Therapeutic plan for PAH

Kaplan-Meier Survival by Age Group

-1 year vs. 1-11 years: p = 0.3215
-1 year vs. 12-17 years: p = 0.7200
1-11 years vs. 12-17 years: p = 0.0330

Survival (%)

1988-1994 vs. 1995-2001: p = 0.0313
1988-1994 vs. 2002-6/2006: p < 0.0001
1995-2001 vs. 2002-6/2006: p = 0.0189

The RV improves within 3 months of lung transplant for PAH


Freedom from Severe Renal Dysfunction*
For Pediatric Lung Recipients (Follow-up: April 1994 - June 2006)

* Severe renal dysfunction = Creatinine > 2.5 mg/dl (221 μmol/L), dialysis or renal transplant

Freedom from Bronchiolitis Obliterans
For Pediatric Lung Recipients (Follow-up: April 1994 - June 2007)

POST-LUNG TRANSPLANT MORBIDITY FOR PEDIATRICS
Cumulative Prevalence in Survivors within 7 Years Post-Transplant
(Follow-up: April 1994 - June 2006)

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Within 7 Years</th>
<th>Total number with known response</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertension</td>
<td>74.5% (N = 55)</td>
<td></td>
</tr>
<tr>
<td>Renal Dysfunction</td>
<td>36.2% (N = 58)</td>
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<tr>
<td>Abnormal Creatinine &gt; 2.5 mg/dl</td>
<td>24.1%</td>
<td></td>
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<tr>
<td>Chronic Dialysis</td>
<td>5.2%</td>
<td></td>
</tr>
<tr>
<td>Renal Transplant</td>
<td>0.6%</td>
<td></td>
</tr>
<tr>
<td>Hyperlipidemia</td>
<td>5.2% (N = 58)</td>
<td></td>
</tr>
<tr>
<td>Diabetes</td>
<td>31.6% (N = 57)</td>
<td></td>
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<tr>
<td>Bronchiolitis Obliterans</td>
<td>28.6% (N = 35)</td>
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</tbody>
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Last updated based on data as of December 2006
Conclusions

- Lung transplantation works as a treatment for end-stage PH.
- The listing process is different for children under vs over 12 y and still does not favor PH patients.
- Longterm results are improving rapidly with a better understanding of the morbidity of previous therapies.

- Evaluate and list appropriately, transplant when ready.

Tom Spray’s additional comments

- Results of lung and heart-lung transplant for PH similar to results for other indications
- Complications / early deaths may be increased due to previous surgeries and diagnoses
- Cyanosis in association with previous thoracotomy may be contraindication
- Lung tx with CHD repair may carry increased risk of death compared to other indications
Conclusion

“Although lung and heart/lung transplantation are imperfect therapies for pulmonary arterial hypertension, when offered to an appropriately selected population transplantation may offer prolongation of an improved quality of life.”