The Indian Subcontinent

- Total population 1.57 billion
- 3 of top 10 countries by population
- 22.7% of world population live here

Demographics of India

<table>
<thead>
<tr>
<th>Population</th>
<th>1.14 billion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Crude birth rate</td>
<td>22.1% in (2008)</td>
</tr>
<tr>
<td>GDP/capita (USD)</td>
<td>1508 in 2000, 3452 in 2008</td>
</tr>
<tr>
<td>GDP rank</td>
<td>(161/225 nations)</td>
</tr>
<tr>
<td>Human development</td>
<td>132/175 available nations</td>
</tr>
<tr>
<td>Index ranking</td>
<td>-</td>
</tr>
<tr>
<td>Population below poverty line</td>
<td>~ 27%</td>
</tr>
</tbody>
</table>

Congenital Heart Disease in India

- Newborns with disease @ 8/1000 live births – 2,00,000 yr
- Newborns with serious heart disease @ 2/1000 live births (most conservative estimate) – 50,000/yr
- No. of cardiac surgery for CHD estimated – 6000 in 2001
- About 10,000 now
- No. of infant cardiac surgeries – 1200 in 1998, 2500 in 2007

Pediatric Cardiologic Priorities in India

Magnitude & scope of problem:
Life-threatening CHD - 2 - 3/1000 live births

1998:
- 40000 - severe CHD
- 1200 operated

2007:
- 60000 - severe CHD
- 2500 operated

Pediatric Cardiac Care in India – A Reality Check

<table>
<thead>
<tr>
<th></th>
<th>India</th>
<th>US</th>
</tr>
</thead>
<tbody>
<tr>
<td>Population</td>
<td>1.1 billion</td>
<td>300 million</td>
</tr>
<tr>
<td>CHD diagnosis</td>
<td>&gt; 96% missed in infancy</td>
<td>In utero</td>
</tr>
<tr>
<td>Ped Cardiologists</td>
<td>50</td>
<td>2500</td>
</tr>
<tr>
<td>Dedicated surgeons</td>
<td>20</td>
<td>200</td>
</tr>
<tr>
<td>Dedicated centers</td>
<td>1 for 100 million population</td>
<td>1 for &lt; 5 million population</td>
</tr>
</tbody>
</table>
Barriers to Pediatric Cardiac Care in India

- **Accessibility**
  - Facilities in major metros
  - No referral system

- **Affordability**
  - Mostly individual spending
  - Mostly private enterprises

- **Acceptability**
  - Female gender
  - Social factors

Etiologies – PAH in Children in India

- IPAH
- Un-operated CHD
- RHD

PAH with Congenital Heart Disease

- 11,500 neonates born in a community hospital
- Echocardiographic screening
- Serious CHD - 39 neonates
  - 3.4/1000 live births
- PPHN - 4 neonates (0.3/1000)
- At risk of severe PAH - 27 neonates (2.3/1000)


Operability Testing in India

- 100 consecutive catheterization for operability (2008 – 10)
- Mean age
  - VSD – 8.5 years
  - PDA – 10.7 years
  - ASD – 31.6 years

Operability Testing In PDA PAH

10 – 25% Cardiac catheterizations are done for operability issues
Operability Testing – Case 1

8-year-old presented with exertional dyspnea for the last 4 years
- His hemodynamic data & angiographic data are shown first - (for keeping up the story)

Catheterization Data

<table>
<thead>
<tr>
<th></th>
<th>mm Hg</th>
<th>Saturation %</th>
</tr>
</thead>
<tbody>
<tr>
<td>RA</td>
<td>7</td>
<td>58</td>
</tr>
<tr>
<td>RV</td>
<td>106 / 7-8</td>
<td>62</td>
</tr>
<tr>
<td>PA</td>
<td>108 / 76</td>
<td>90</td>
</tr>
<tr>
<td>AO</td>
<td>116 / 76</td>
<td>92</td>
</tr>
<tr>
<td>PVRI(U)</td>
<td>66 (FA)</td>
<td></td>
</tr>
</tbody>
</table>

Pre-op chest x-ray

Operability Testing – Case 2

11-years-old male
- Transposition of great arteries – VSD PAH
- Catheterized at 2 years of age

Catheterization Data at 2 years of age

<table>
<thead>
<tr>
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<th>mm Hg</th>
<th>Saturation %</th>
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<tbody>
<tr>
<td>RA</td>
<td>5</td>
<td>54</td>
</tr>
<tr>
<td>RV</td>
<td>100 / 5 – 6</td>
<td>62</td>
</tr>
<tr>
<td>PA</td>
<td>106 / 50</td>
<td>70</td>
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<tr>
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Advised palliative Senning
Operability issues in CHD

- Operability in older children and adults with CHD is a relevant question in many parts of the world
- Despite advances in understanding of CHD, this issue remains an ‘inexact science’

PAH with CHD in Indian Subcontinent
Eisenmenger Syndrome in India

**Eisenmenger Syndrome in India**

The AIMMS registry
- 50 patients of Eisenmenger syndrome
- Mean age 24.4 ± 10.2 years
- Followed up for a mean duration of 18.2 months
- Clinical events
  - Death – 2 (4%)
  - Worsening HF – 5 (10%)

38% had hsCRP > 3 mg/L
IL-2, IL-6, TNF not elevated

Predictors of clinical worsening: 6mwd & HR following 6 min walk

**Operability Testing – Case 2**

Catheterization Data at 11 years of age

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Catheterization 3 months post ASO & Double patch VSD Closure

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<td>38 / 7- 8</td>
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<td>38 / 12 23</td>
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Beta-blockers in ES

- 15 patients with ES – Acute hemodynamics & safety of metaprolol (iv & oral for 6 weeks)

Baseline 6 Weeks

Operability Testing – Case 2

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PAH in Rheumatic Heart Disease

RHD in India - Is it Declining?

Prevalence (per 1000)

1972-75
1984-87
2002-05

0
1
2
3
4
5
6

ICMR School Surveys

Author
Shrestha et al. 1991
Regmi et al. 1997
ManBahadur et al. 2002
Rizvi et al. 1993
Mendiset al. 1998

Region
Kathmandu
Kathmandu
Rural
Rural

Popn.
4,816
4984
7,188
9483

Age (yrs)
5 to 16
5 to 16
6-18

Prev (/1000)
1.35
1.2
0.9

RHD Decline – Is it ‘Real’?

Prevalence is still high amongst poor and the underprivileged

RHD may have declined among the privileged 25% of the Indian population

Burden may Not Decline

<table>
<thead>
<tr>
<th>Year</th>
<th>Total Population</th>
<th>Prevalence (%)</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1981</td>
<td>685</td>
<td>1.52</td>
<td>1.04</td>
</tr>
<tr>
<td>1991</td>
<td>846</td>
<td>0.29</td>
<td>1.59</td>
</tr>
<tr>
<td>2001</td>
<td>1027</td>
<td>0.21</td>
<td>1.4</td>
</tr>
<tr>
<td>2006</td>
<td>1130</td>
<td>0.04</td>
<td>0.44</td>
</tr>
</tbody>
</table>
Echocardiographic prevalence of RHD

RHD – A Treatable Cause of PAH

Acute Hemodynamic Results of PTMC

Event Free Survival following PTMC

PTMC in Children

**PTMC below 12 yrs – ALLMS experience**

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>Mean ± SD</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>11 ± 1.18</td>
<td>(7 - 12)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Wt. (Kg)</th>
<th>Mean ± SD</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>23.5 ± 6.4</td>
<td>(11.3 - 40)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Ht. (Cm)</th>
<th>Mean ± SD</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>137.1 ± 12.1</td>
<td>(110 - 156)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>NYHA III / IV</th>
<th>Mean ± SD</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>68 / 12 (80%)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Success rates:
- PTMC: 80%
- Early PTMC: 100%
- Late PTMC: 50%

**Conclusions**
- PTMC in young children is safe & effective
- Cautious, stepwise Balloon Dil. required
- Acceptable risks of MR, Restenosis
- Palliative procedure
Idiopathic PAH in Children

AIIMS Data 2009-10

- 14 patients
- Mean age 9.92 ± 3.99 yrs (6 months – 14 years)
- Male 7, female 7
- At presentation
  - Syncope: 7/14 (50%)
  - Congestive failure: 8/14 (57%)
  - RVSP by echo: 74.92 ± 15.66 mm Hg
  - RV dysfunction: 12/14 (86%)

Children often present late with advanced disease

PAH Therapies & Cost

<table>
<thead>
<tr>
<th>Drug</th>
<th>Cost per month ($)</th>
<th>Cost / Average per capita income</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sildenafil</td>
<td>7</td>
<td>0.1</td>
</tr>
<tr>
<td>Bosentan</td>
<td>120</td>
<td>1.3</td>
</tr>
<tr>
<td>Inhaled prostacycline</td>
<td>2960</td>
<td>32.5</td>
</tr>
<tr>
<td>Intravenous Prostacycline</td>
<td>5000</td>
<td>54.1</td>
</tr>
</tbody>
</table>

CTEPH in India

- Thrombo-embolic PAH thought to be less common in Asians
- Factor V Leiden less common
- VTE is no longer a rarity in India (Lee et al 2009)
- PTE remains undiagnosed

Politics is nothing more than medicine on a Grand Scale
Virchow, 1860

Medicine is nothing more than politics on a Grand Scale...
PAH in Children – Absent RPA

PAH in Children – Absent RPA

PAH in Children – Absent Pulmonary Veins

Posterior view showing RPA occlusion & distal RPA

PA-120/70 (95)
FA - 85%

$\frac{1}{R} = \frac{1}{R_1} + \frac{1}{R_2}$
Pulmonary veno-occlusive disease


Dietary factors in PVOD & other forms of PAH?

Kothari SS et al. Circulation 2009

Pulmonary capillary Hemangiomatosis

Neonatal Pulmonary Hypertension - ? PPHN

Author Location/ Tribe n Prevalence (%) PAH

\begin{tabular}{|c|c|c|c|}
\hline
Author & Location/ Tribe & n & Prevalence of PAH for India \%
\hline
Deshmukh et al & Maharashtra & 5561 & 2.9
\hline
Mandot et al & Rajasthan & 1676 & 9.2
\hline
Vasava et al & Gujarat & 948 & 25.5
\hline
Feroze et al & Kerala & 714 & 24.6
\hline
Kamble et al & Maharashtra & 1753 & 5.7
\hline
\end{tabular}

PAH in India – Sickle Cell Anemia

PAH in India – Filariasis

\begin{itemize}
\item 23 million cases of symptomatic filariasis
\item Unlike schistosomiasis, not associated with PAH
\end{itemize}

PAH in India - Endomyocardial Fibrosis (EMF)
PAH in India - Takayasu Arteritis

Kothari SS, Images Paediatr Cardiol 2002

PAH in India

- Burden remains unknown
- Diagnosis is often delayed
- Outcome may be less than satisfactory

Intellectually challenging
Emotionally draining

An Obstinate Patient

Kothari SS, Br Med J 2004

O’ God, Grant me
Serenity to accept the things
I can not change,
Courage to change
The things I can,
And, Wisdom
To know the difference.

Clairvoyance
humanitiesweb.org