Long-term Functional Health Status and Exercise Test Variables for Patients with Pulmonary Atresia with Intact Ventricular Septum: A Congenital Heart Surgeons’ Society Study

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Disclosures

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Case

Female neonate

- PAIVS
- TV Z-score -2
- RV sinusoids
- No RV-dependent circulation

What would you do?

- UV, 1.5 V, BV
Repair, TV, and Survival

![Graph showing the relationship between tricuspid valve Z-score and survival outcomes.]

- **Univentricular**
- **Biventricular**
- **Death**

1.5 V
Choice of repair strategy and TV Z score ...how are they related to long-term functional health status and exercise capacity?
Outline

1987 – 1997
N=448 neonates
PAIVS

Prospective evaluation:
- Perceived functional health status
- Exercise capacity
Participants

271 survivors from initial cohort

106 participants (39%)

- UV (N=25)
- 1.5V (N=14)
- BV (N=63)
How do they feel?
Psychosocial and Emotional Domains

Domain Score

Normals
PAIVS

*
Repair strategy, TV, and FHS

Only 1 of 23 domains showed any difference
  • Global general health (CHQ)

No influence of repair strategy on FHS

No influence of TV Z-score on FHS
How well can they exercise?
Tricuspid Valve Z-Score
Peak VO2 and TV Size

Peak Oxygen Consumption (% predicted)

Initial Tricuspid Valve Z-Score
Peak VO2, TV Size, and Repair Pathway

Peak Oxygen Consumption (% predicted)

Initial Tricuspid Valve Z-Score
Case

Female neonate

- PAIVS
- TV Z-score -2
- RV sinusoids
- No RV-dependent circulation

What would you do?

- UV, 1.5 V, BV
...19 years later

How will she feel?
- Worse than normal physical functioning
- Better than normal perceived FHS

How will she exercise?
- 1V - 82%
- 1.5V – 90%
- 2V – 765
Conclusions
Late FHS

Reduced regardless of repair pathway compared to normals

Not related to the initial TV Z

Dichotomy – patients with PAIVS feel as if they are doing well despite physical impediments
Late Exercise Capacity

Reduced regardless of repair pathway compared to normal

Borderline patients with smaller initial TV Z may be better served with 1.5 V or UV

TV Z clinically useful for predicting late aerobic capacity