Successful Transition of the Late Adolescent PH Patient: Canadian Transition Program

Presented by:

Janette T. Reyes, MN, NP-Peds
Nurse Practitioner
Pulmonary Hypertension Service,
The Labatt Family Heart Centre
The Hospital for Sick Children, Toronto, CANADA

Premise

- Survival in children with Pulmonary Hypertension (PH) has improved since the 1990s:
  - new targeted medical therapies
  - medical advances

Premise

<table>
<thead>
<tr>
<th>Survival of IPAH/familial PAH at 1, 3, 5, 7 years from time of diagnosis:</th>
</tr>
</thead>
<tbody>
<tr>
<td>REVEAL registry (2006-2009)</td>
</tr>
<tr>
<td>NIH registry (1981-1988)</td>
</tr>
</tbody>
</table>

Premise

<table>
<thead>
<tr>
<th>Survival pediatric IPAH/familial patients enrolled from time of diagnosis:</th>
</tr>
</thead>
<tbody>
<tr>
<td>REVEAL registry (2006-2009)</td>
</tr>
</tbody>
</table>

Premise

- Increase in pediatric PH patients surviving into adulthood
- Moving to adult care during a vulnerable period of development with adolescents – process should be dealt with in a sensitive manner
Aim of Health Care Team

To execute a smooth transition and provide a positive transfer to adult care experience, for the adolescent patient diagnosed with Pulmonary Hypertension.

Goal of Patient

- be empowered
- take responsibility for own health and lifestyle
- attain best quality of life

Transition Process

- Role of the pediatric health care team:
  - begin preparation at an early age
  - discuss transition plan and expectations
  - involve and support parent(s)/guardian(s)
  - comply with institution policy

- Role of the adult health care team:
  - involvement of family for first few visits
  - continue with the transition as it reaches it end
  - comply with institution policy

Adolescent Transition Process: Information Discussed

- diagnosis, treatment - communication style
- medications
- psychosocial issues and feelings about transfer
- changes in insurance coverage
- show how to access resources
- provide information about Adult PH Centre
Adolescent Transition Process: Information Discussed

- Sexuality
- Nutrition
- Dental care
- Lifestyle
- School
- Employment
- House chores

Information for Patients with Developmental Challenges and Needs

Link with:

- Developmental Services agencies
- Ontario Disability Support Program (apply 6 months prior to 18th birthday)
- Assistive Devices Program

Transition Process at the Pediatric Centre: Helpful Aids

- Passport for wallet/purse – meds, appointments, doctors’ address and telephone numbers
- Medical Alert Bracelet
- Resource Websites
- Support Groups
**The Hospital for Sick Children: Transition of Patients**

- **PH Transition Workshop (established 2007):**
  1. Group - x4 hours, completed on a set date
  2. Individually - completed at last clinic appointment

- In collaboration of Good 2 Go Transition Program (established 2006)

**The Hospital for Sick Children: PH Transition Workshop Agenda**

- Meet and Greet – Overview, Checklist completion
- Good to Go Transition Program Presentation
- Adult PH Program - Introduction of Adult team
- Break Out Session – Parents’ room; Teens’ room
- My Health Passport
- Tour at Adult PH Centre
- Graduation/Evaluation

**Evaluation**

**Pre Workshop:**
Nervousness: Caregivers more than patients

- **Caregivers:**
  - Level of care will change
  - If same attention to patient
  - Unfamiliarity of hospital and staff

- **Patients:**
  - Having a new NP and Doctor
  - Not knowing anyone
  - Big change

**Post Workshop:**
Nervousness: Caregivers decreased, patients increased

- **Caregivers:**
  - Tour
  - Meeting the doctor
  - 100% workshop helpful

- **Patients:**
  - Tour
  - “everything”
  - 90% workshop helpful
Challenges: Transition Workshop Day
- Patients long travel to hospital
- Finding one day where patients, families, Transition Team, Adult Hospital Caregivers available
- Lack of effect with patients with Trisomy 21 or developmental challenged
- Varying length of follow-up in Clinic – some only seen once per year

Challenges: Pre Transfer
- Patients and families request transfer to particular PH Center of Excellence
- Patients and families unable to attend Transition Workshop Day
- Distance walked during tour

Challenges: Post Transfer
- Medical records not transferred in timely manner
- Pediatric Center unaware of delay in transfer of medical records
- Missing information of medical records

Conclusion
Successful transition in the adolescent is possible
- Start with a transition plan.
- Necessary continuation of:
  - Collaboration - health team members in pediatric and adult centre of excellence
  - Evaluation - young adolescent’s experience pre and post transition
References

- Hudsmith, LE and Thorne, SA. Transition of care from paediatric to adult services in cardiology. Archives of Disease in Childhood. 2007;92, 927-930.

Disclosures

- United Therapeutics Advisory Board
- Actelion Advisory Board
Objectives

• Substantiate need for formal transition program.

• Describe obstacles to transition program within the United States healthcare system.

• Propose transition program model

Natural History of IPAH
NIH Registry\textsuperscript{1,2}

\begin{itemize}
  \item Predicted survival according to the NIH equation. Predicted survival rates were 69\%, 56\%, 46\%, and 38\% at 1, 2, 3, and 4 years, respectively.
  \item The numbers of patients at risk were 231, 149, 82, and 10 at 1, 2, 3, and 4 years, respectively.
  \item Patients with primary pulmonary hypertension, now referred to as idiopathic pulmonary hypertension.
\end{itemize}


Acute to Targeted Therapy:

\begin{itemize}
  \item CCB, anticoagulation, digitalis, diuretics
  \item Epoprostenol
  \item SC treprostinil
  \item IV treprostinil
  \item Iloprost
  \item Sildenafil
  \item Ambrisentan
  \item Tadalafil
  \item Inhaled Treprostinil
\end{itemize}

Pediatric Survival from Diagnosis: REVEAL

### Pediatric Survival from Diagnosis: UK

![Graph showing survival rates for pediatric PAH patients in the UK.](image)


### Survival of Pediatric PAH in Combined Netherlands Cohorts: 1991 - 2005

![Graph showing survival rates for pediatric PAH patients in the Netherlands.](image)


### A Fortunate Dilemma: Bridging the Gap

- **Pediatric Model**
  - Family Centered
  - Dependent on Parent
  - Family Oversight
  - CHD Associated PH

- **Adult Model**
  - Patient-Centered
  - Independence
  - Individual Accountability
  - Autonomy

### Different Healthcare Systems, Different Challenges

- **Government Supported Medicine**
  - Center of excellence for country or region
  - PH patients transition to fewer Adult Program
  - Patients may live closer to PH Center

- **Private Supported Medicine**
  - Few Pediatric Centers
  - Many Adult Centers
  - Large geography of US
  - Many patients live out-of-state many miles from Pediatric PH Center
Collaborative Transition = Successful Transfer

- Transition Program Barriers
  - Insufficient staffing
  - No identified staff members responsible for transitions
  - Financial challenges
  - Institutional acceptance
  - Resistance from adolescent and parents
  - Hesitancy of pediatric provider.

- Key Transition Concepts:
  - Timing
  - Patient, Family, and Provider readiness
  - Identification of adult PH care team
  - Buy-in from pediatric and adult programs
  - Successful completion of transition curriculum
  - Transfer of care

Transition Models

- Society of Adolescent Medicine 1993 Position Paper
- Adult Congenital Heart Disease 2011 Best Practices Statement
- Cystic Fibrosis Foundation

References:
Children's Hospital Colorado (CHCO) PH Program Transition Roadmap

**PHASE III**

17-18 years

Curriculum:
- Discuss high risk behaviors
- Females: Discuss risks of pregnancy to mother and fetus
- Discuss safe contraceptive options
- Discuss education/employment plan
- Discuss health insurance options
- IV/SQ/Inhaled Prostacyclin Patient: Fully competent with IV or SQ infusion or inhaled prostacyclin administration

Roles/Responsibilities:
- Patient: Discuss thoughts/concerns of transfer readiness.
- Parent: Review with patient gaps in skills. Discuss thoughts/concerns of transfer readiness.
- Provider: Assist family to identify Adult PH program options. Be understanding of patient/parent's perspective.
- Communicate with Adult PH program regarding transfer referral.

Successful completion of PHASE III curriculum

**PHASE IV**

18 - 21 years

Transfer of Care

Follow-up phone conference between providers

Successful Transfer

Roles/Responsibilities:
- Patient: Review and demonstrate competency of self-care management with pediatric care team.
- Parent: Review healthcare coverage with insurance provider and discuss with adolescent.
- Pediatric Provider: Send patient transfer referral packet to Adult PH care provider.
- Adult Provider: Accept care and responsibility for PH patient.

*Refer to Transition Completion checklist for additional provider responsibilities

Transfer Checklist

- Patient and family and pediatric PH provider determine readiness to transition.
- Patient has successfully completed Phases I through III of transition curriculum.
- Adult PH provider identified and notified of transfer referral.
- Adult PH provider has accepted referral and has received transfer referral packet.
- Patient and parents have the Adult PH program's contact information.
- Medical history and genetic testing results are up to date.
- Patient has scheduled and completed first adult PH clinic visit.
- Follow-up care conference between pediatric and adult providers to discuss issues.
- Patient transfer of care complete and hand off successful.
- Patient has contact with pharmaceutical, home health, and equipment providers with new PH provider information for future prescription management and care orders.

Conclusion

- Dialogue
  - Adult Programs
  - PH Community
  - Pediatric Colleagues
- Commitment to Collaboration from Adult Colleagues
- Evaluation and Feedback Loop
  - Young Adults after Transition
  - Adult Colleagues
- Advocacy and Support from Pulmonary Hypertension Association and Pediatric Support Groups.
Pulmonary Hypertension Program
Children's Hospital Colorado