Adaptation and Resilience in Families of Individuals with Down Syndrome

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  - School of Nursing

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  - Assistant Professor
  - Kyungpook National University
  - College of Nursing

Down Syndrome

- Most common genetic cause of intellectual disabilities
  - 1 in every 800-1000 live births
  - 1 in 691 in the United States
- Complex Disorder
  - Multigene/Multisystem
- High degree of variability

Worldwide,

- There are over 7 million people living with Down syndrome
- Families of individuals with Down syndrome are from all races, religions, ethnic backgrounds and socioeconomic status
Due to advances in health care, people with DS are living longer, more fulfilling lives

- Average life expectancy for individuals with DS was 9 years in the early 1900’s
- Now it is around 60 (in developed countries)
  - Bert Holbrook - age 80

However, a great deal of stigma still exists

- In many countries, families face daily challenges related to inaccurate, outdated ideas about Down syndrome
  - Termination rates are as high as 98% in some countries
- Services & resources vary greatly from one country to the next

Access to prenatal testing also varies greatly from one country to the next

- In September 2004 the Danish National Board of Health issued new guidelines for prenatal screening and diagnosis
- Dramatic decrease in number of infants born with Downs syndrome
  - 55-65 per year 2000-2004
  - 31 in 2005
  - 32 in 2006
- Speculation - Denmark will become “Down Syndrome Free Society” by 2030

Outline

- Overview of Main Study
- Preliminary Findings (findings from 4 countries)
  - Maternal Satisfaction with Family-Provider Interactions Surrounding the Diagnosis of Down Syndrome
  - Influence of Culture and Family Factors on Adaptation in Families of Individuals with Down Syndrome

Main Study

- Purpose
  - To examine how culture, interactions with health care providers and family factors contribute to adaptation and resilience in families of individuals with Down Syndrome
**Approach: Research Methods & Design**

- Cross-Cultural (10 or more countries)
- Cross-Sectional, Descriptive, Mixed Methods
  - All participants complete a packet of self-report questionnaires designed to assess key dimensions of the Resiliency Model of Family Stress, Adjustment & Adaptation
    - Hard copy or Online version
    - Participants are given the option of being interviewed
      - In-person or Telephone

**Family Measures**

<table>
<thead>
<tr>
<th>Measure</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Family Index of Regenerativity and Adaptation (FIRA-G)</td>
<td>74 item measure/7 scales</td>
</tr>
<tr>
<td>Family Management Measure (FaMM)</td>
<td>53 item measure/6 scales</td>
</tr>
<tr>
<td>Family Problem Solving Communication Index (FPSC)</td>
<td>10 item measure</td>
</tr>
<tr>
<td>Family Member Well-Being (FMWB)</td>
<td>8 item measure</td>
</tr>
<tr>
<td>Brief Family Assessment Measure General (BFAM-G)</td>
<td>14 item measure</td>
</tr>
<tr>
<td>Background Information</td>
<td>Demographic Information Questions about how they were informed of diagnosis of Down Syndrome</td>
</tr>
</tbody>
</table>

**Family Variables/Measures**

<table>
<thead>
<tr>
<th>Measure</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Family Demands</td>
<td>Family Stress Index (FSS) FIRA-G</td>
</tr>
<tr>
<td></td>
<td>Condition Management Effort Scale (4 items FaMM)</td>
</tr>
<tr>
<td>Family Appraisal</td>
<td>Child’s Daily Life (5 item FaMM)</td>
</tr>
<tr>
<td></td>
<td>Condition Management Ability (2 item FaMM)</td>
</tr>
<tr>
<td></td>
<td>Family Life Difficulty (14 items FaMM)</td>
</tr>
<tr>
<td></td>
<td>View of Condition Impact Tool (10 items FaMM)</td>
</tr>
<tr>
<td></td>
<td>Parental Morality Scale (6 items FaMM)</td>
</tr>
<tr>
<td>Family Type</td>
<td>Family Hardiness Index (20 items FIRA-G)</td>
</tr>
<tr>
<td>Family Resources</td>
<td>Relative and Family Support (8 items FIRA-G)</td>
</tr>
<tr>
<td></td>
<td>Social Support Index (17 items FIRA-G)</td>
</tr>
<tr>
<td>Family Problem Solving &amp; Coping</td>
<td>Family Problem-Solving Communication Index (12 items FaMM)</td>
</tr>
<tr>
<td></td>
<td>Family Coping-Cohesion (6 items FIRA-G)</td>
</tr>
<tr>
<td>Family Adaptation</td>
<td>Brief Family Assessment Measures (14 item B-FAM)</td>
</tr>
<tr>
<td></td>
<td>Family Distress (5 items FIRA-G)</td>
</tr>
<tr>
<td></td>
<td>Family Member Well-Being (8 items FMWB)</td>
</tr>
</tbody>
</table>

**Resiliency Model of Family Stress, Adjustment, and Adaptation**

**Progress to Date**

- Over 1200 parents have completed the self-report questionnaires (Brazil, Ireland, Japan, Korea, Malaysia, Netherlands, Portugal, Thailand, United Kingdom, United States)
- Over 600 parents have expressed interest in being interviewed
- Data collection will begin in other countries this spring (Italy, Spain, hopefully Sweden and Denmark)
Maternal Satisfaction with Family-Provider Interactions Surrounding the Diagnosis of Down Syndrome

What do we know about the Informing Process?

- Numerous studies concerning parental satisfaction with informing process
- Many articles about how to break “bad or difficult news” to patients and families
  - Common exemplar – unexpected diagnosis of DS
- A number of guidelines and educational resources to help health care providers feel better prepared to deliver diagnosis of DS

Unfortunately,

- There continue to be reports of:
  - Health care providers giving parents inaccurate, out-dated information about life with DS
  - Parents feeling pushed or coerced to make unwanted choices, such as undergoing invasive testing or terminating a pregnancy following the diagnosis of DS

Aim of Study

- To explore maternal satisfaction with family-provider interactions surrounding the diagnosis of Down syndrome.

For this study

- Focus was limited to responses from 710 mothers living in:
  - Ireland (100)
  - Portugal (68)
  - USA (451)
  - United Kingdom (91)
### Initial Awareness of Diagnosis

<table>
<thead>
<tr>
<th></th>
<th>Before</th>
<th>Other</th>
<th>After</th>
</tr>
</thead>
<tbody>
<tr>
<td>All Mothers</td>
<td>14.47%</td>
<td>10.50%</td>
<td>75.04%</td>
</tr>
<tr>
<td>Ireland</td>
<td>12.75%</td>
<td>9.80%</td>
<td>77.45%</td>
</tr>
<tr>
<td>Portugal</td>
<td>5.88%</td>
<td>8.82%</td>
<td>85.29%</td>
</tr>
<tr>
<td>United Kingdom</td>
<td>2.35%</td>
<td>10.59%</td>
<td>87.06%</td>
</tr>
<tr>
<td>USA</td>
<td>18.44%</td>
<td>10.89%</td>
<td>70.67%</td>
</tr>
</tbody>
</table>

### Parental Satisfaction with Informing Process

- Continues to be less than ideal
- However, many are satisfied - especially those who were given
  - Up-to-date, accurate information
  - Information about what life is like for individuals with DS and their families

### Satisfaction with how informed

<table>
<thead>
<tr>
<th></th>
<th>Percent of Mothers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Very Satisfied</td>
<td>22.82%</td>
</tr>
<tr>
<td>Moderately Satisfied</td>
<td>22.96%</td>
</tr>
<tr>
<td>Slightly Satisfied</td>
<td>9.86%</td>
</tr>
<tr>
<td>SATISFIED</td>
<td>55.64%</td>
</tr>
<tr>
<td>Slightly Dissatisfied</td>
<td>9.44%</td>
</tr>
<tr>
<td>Moderately Dissatisfied</td>
<td>8.59%</td>
</tr>
<tr>
<td>Slightly Dissatisfied</td>
<td>26.34%</td>
</tr>
<tr>
<td>DISSATISFIED</td>
<td>44.37%</td>
</tr>
</tbody>
</table>

### Percent Satisfaction by Country

<table>
<thead>
<tr>
<th></th>
<th>Ireland</th>
<th>Portugal</th>
<th>United Kingdom</th>
<th>United States</th>
</tr>
</thead>
<tbody>
<tr>
<td>Very Sat</td>
<td>27.00%</td>
<td>20.59%</td>
<td>31.87%</td>
<td>20.40%</td>
</tr>
<tr>
<td>Mod Sat</td>
<td>27.00%</td>
<td>30.88%</td>
<td>24.18%</td>
<td>20.62%</td>
</tr>
<tr>
<td>Sl Sat</td>
<td>8.00%</td>
<td>20.59%</td>
<td>6.59%</td>
<td>9.31%</td>
</tr>
<tr>
<td>SAT</td>
<td>62.00%</td>
<td>72.06%</td>
<td>62.64%</td>
<td>50.33%</td>
</tr>
<tr>
<td>Sl Dis</td>
<td>10.00%</td>
<td>2.94%</td>
<td>8.79%</td>
<td>10.42%</td>
</tr>
<tr>
<td>Mod Dis</td>
<td>6.00%</td>
<td>5.88%</td>
<td>9.89%</td>
<td>9.31%</td>
</tr>
<tr>
<td>Sl Dis</td>
<td>22.00%</td>
<td>19.12%</td>
<td>18.88%</td>
<td>29.93%</td>
</tr>
<tr>
<td>DIS-SAT</td>
<td>38.00%</td>
<td>27.94%</td>
<td>37.36%</td>
<td>49.66%</td>
</tr>
</tbody>
</table>

### Predicting Satisfied vs Not Satisfied using selected predictors (Mothers)

- Only predictor that was significant was country:
  - Mothers from Portugal were significantly more likely than mothers from US, UK, and Ireland to be satisfied with how they were informed of child’s diagnosis (P=0.009).
- Predictors that were non significant include:
  - Who informed them
  - Child’s age
  - When diagnosis was made

### Improvement overtime? No, change was not significant

- Relationship between age of the child with DS and satisfaction with how they were informed was nonsignificant (P=0.976)
- Parents of younger children with DS did not report significantly higher levels of satisfaction with how they were informed
**Difference in when diagnosed? No**

- Relationship between timing of diagnosis and satisfaction with how informed was nonsignificant

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**Two open-ended questions**

1. Please indicate at least one thing that health care providers did or said that you found helpful?
2. Please identify at least one thing that you would have liked health care professionals to have done differently?

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**Fairly close match between parent preferences & recent guidelines**

- Parents wanted:
  - To be told together in a joint meeting with a health care provider they knew and trusted.
  - The conversation to take place in a private setting as soon as possible after the diagnosis of DS was suspected.
  - Health care providers to be upfront, honest, and forthcoming with accurate, up-to-date information

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**What parents found most helpful**

1. Unbiased attitude of health care providers (e.g., presented balanced information, did not try to influence parental decisions)
2. Positive comments about the child and what the child will most likely be able to accomplish
3. Emotional support (e.g., caring, sensitivity, & hope)
4. Material support (e.g., provision of up-to-date resources/information about local support groups)
5. Provision of information in a timely manner

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**Specific Responses**

- One mother noted, “they need to present the information gently and personally – not a sterile statement void of any human emotion”.
- Another indicated that she wished providers had not acted as if they were delivering “bad news”.
- A father wrote, “I wish we had known right from the start what a positive experience we could have with a child who has Down Syndrome”

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**Parents wanted more information about family experience of living with DS**

- Potential impact on:
  - Siblings
  - Parental relationship
  - Family as a whole
Possible explanation for why many providers fail to provide information about the family experience of DS

- Health care providers who deliver the diagnosis of DS are not usually the same providers who provide ongoing care to children with DS and their families
- Their views about the family experience of DS are often based on the initial responses of parents/families to the diagnosis of DS

This is problematic because

- In most families of individuals with DS, views of the situation change dramatically over time.

Initial vs Current View - Mothers

<table>
<thead>
<tr>
<th></th>
<th>Initial</th>
<th>Current</th>
</tr>
</thead>
<tbody>
<tr>
<td>A blessing in disguise</td>
<td>8.71%</td>
<td>59.49%</td>
</tr>
<tr>
<td>Challenge to be overcome</td>
<td>27.67%</td>
<td>16.17%</td>
</tr>
<tr>
<td>Just something to accept</td>
<td>20.08%</td>
<td>23.35%</td>
</tr>
<tr>
<td>A tragedy</td>
<td>24.16%</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>The worst thing that could have happened</td>
<td>19.38%</td>
<td>&lt;1%</td>
</tr>
</tbody>
</table>

“...It is really not quite the tragedy... At the time you really feel that this is the biggest tragedy that ever happened. If we could have known what it would be like to have M., we wouldn’t have been nearly so sad. No one really mentioned the positive side.”

Conclusion – for this study

- Parental preferences concerning the informing process match fairly well with current guidelines
- The information expectant and new parents receive about DS needs to be:
  - Up-to-date and accurate
  - Provided in a timely manner
  - And it needs to include information about the family experience of living with DS

Families are likely to benefit by hearing that negative consequences are not inevitable – in fact many families not only survive – they thrive!
Cross-Cultural Study of Adaptation in Families of Individuals with Down Syndrome

For this study

- Data from 802 parents living in 4 countries:
  - Ireland (79 Mothers/18 Fathers)
  - Portugal (63 Mothers/10 Fathers)
  - USA (425 Mothers/119 Fathers)
  - United Kingdom (77 Mothers/11 Fathers)

Approach Used to Analyze Data

- Linear Mixed Modeling
  - 13 Predictors
  - 3 Outcomes
  - All models accounted for:
    - Gender of parent
    - Partner status (partnered or non-partnered)
    - Country
    - Intra-familial correlation and constant variance in families where 2 parents participated

13 Resiliency Model Predictors

<table>
<thead>
<tr>
<th>Predictor</th>
<th>Mothers (644)</th>
<th>Fathers (158)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Family Stressors (higher more stressors)</td>
<td>9.6 (0-40.2)</td>
<td>8.8 (0-41.7)</td>
</tr>
<tr>
<td>Family Strains (higher more strained)</td>
<td>9.1 (0-41.8)</td>
<td>7.7 (0-41.8)</td>
</tr>
<tr>
<td>Cond Management Ability (higher more manageable)</td>
<td>46.1 (26-60)</td>
<td>46.3 (28-60)</td>
</tr>
<tr>
<td>Child Daili Life (higher more normal life)</td>
<td>17.0 (5-25)</td>
<td>17.3 (8-25)</td>
</tr>
<tr>
<td>Family Life Difficulty (higher more difficulty managing)</td>
<td>30.6 (14-67)</td>
<td>31 (14-61)</td>
</tr>
<tr>
<td>View of Condition Impact (higher greater concern)</td>
<td>26.1 (10-44.4)</td>
<td>26.7 (12-39)</td>
</tr>
<tr>
<td>Cond Management Effort (higher more effort)</td>
<td>11.7 (4-20)</td>
<td>11.5 (4-20)</td>
</tr>
<tr>
<td>Family Hardiness (higher more hardy)</td>
<td>63.9 (37-78)</td>
<td>63.8 (39-78)</td>
</tr>
<tr>
<td>Relative &amp; Family Support (higher more support)</td>
<td>25.9 (8-40)</td>
<td>25.5 (8-37)</td>
</tr>
<tr>
<td>Affirming Communication (higher more affirming communication)</td>
<td>11.5 (0-15)</td>
<td>11.2 (2-15)</td>
</tr>
<tr>
<td>Incendiary Communication (higher more incendiary communication)</td>
<td>4.4 (0-13.8)</td>
<td>4.3 (0-14)</td>
</tr>
<tr>
<td>Family Coping (higher - better coping)</td>
<td>15.7 (8-20)</td>
<td>15.5 (8-20)</td>
</tr>
</tbody>
</table>

3 Outcome Variables

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Mothers (644)</th>
<th>Fathers (158)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adjusted Family Functioning</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• &lt;35 Excellent</td>
<td>45.2 (24-108)</td>
<td>45.4 (20-90)</td>
</tr>
<tr>
<td>• 36-64 Increasing Strengths</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• 45-80 Average</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• 81-108 Increasing Problems</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Family Distress (higher more distress)</td>
<td>4.8 (0-33.6)</td>
<td>4.7 (0-27)</td>
</tr>
<tr>
<td>Family Member Well-Being (higher - better well-being)</td>
<td>47.8 (5-78)</td>
<td>49.3 (8-74)</td>
</tr>
</tbody>
</table>

Results

- Each of the 13 predictors had a significant effect on each of the three outcomes (and the direction of the effect was as predicted)

- Adaptive Modeling was then used to reduce the model with all 13 predictors holding the predictors and the intercept in the model
**Family Functioning**

- Family functioning was worse with:
  - Greater Family Strains
  - More Incendiary Communication
  - Less Affirmative Communication
  - Lower Condition Management Ability
  - Lower Hardiness
- Family Functioning was worse in non-partnered families (p=0.001)
- Fathers were not significantly different from mothers in terms of family functioning

**Family Functioning (continued)**

- Parents from Ireland (p=0.577) and the UK (p=0.412) were not significantly different from parents in the US in terms of family functioning.
- However, parents from Portugal were significantly different from parents in the US.
  - Parents from Portugal had lower family functioning (p=0.001)
- Family Functioning changed significantly with child age (family functioning was worse in families of older children) (p=0.008)

**Parent Well-Being**

- Parent Well-Being was worse with:
  - Greater Condition Management Effort
  - Greater Family Strains and Stressors
  - More Incendiary Communication
  - Lower Condition Management Ability
  - Lower Hardiness
- Fathers were not significantly different from Mothers in terms of Parent Well-Being (p=0.722)

**Parent Well Being Continued**

- Parents from Ireland (p=0.335), UK (p=0.343), and Portugal (p=0.428) were not significantly different from parents in the US in terms of Parent Well-Being.
- Parent Well Being did not change significantly with age (p=0.562)
- Non-partnered parents were not significantly different from partnered parents in terms of Parent Well-Being (p=0.106)

**Family Distress**

- Family Distress was greater with:
  - Greater family strains
  - More incendiary communication
- Families from Ireland (p=0.134), UK (p=0.372), and Portugal (p=0.792) were not significantly different from US families
- Fathers were not significantly different from Mothers in terms of Family Functioning (p=0.357)

**Family Distress Continued**

- Families with non-partnered parents were significantly worse than families with partnered parents in terms of Family Distress (p=0.001)
- Family distress did not change significantly with child’s age (p=0.214)
**Summary: Family Functioning**
- FF was worse with:
  - Greater Family Strains
  - More Incendiary Communication
  - Less Affirmative Communication
  - Lower Condition Management Ability
  - Lower Hardiness

**Summary: Parent Well-Being**
- FF was worse for
  - Families in Portugal
  - Non-Partnered Families
  - Families with Older Individuals with DS

**Summary – Family Distress**
- Family Distress was worse with:
  - Greater family strains
  - More incendiary communication

**Resiliency Model of Family Stress, Adjustment, and Adaptation**

**Family Nursing Colleagues/Collaborators**
- Ireland
  - Maria Caples
  - Caroline Dalton
  - Lynne Marsh
  - Anne-Marie Martin

- Portugal
  - Maria do Ceu Barbieri-Figueiredo
  - Maria Joao Silva
  - Aline Soares dos Santos