A DIME AMONG PENNIES

THE PSYCHO-SOCIAL ADAPTATION OF WOMEN WITH TURNER SYNDROME
The International Turner Syndrome Conference:
University of Copenhagen 27 - 30 August 2009
AN AUSTRALASIAN STUDY OF 117 WOMEN WITH TURNER SYNDROME

Dr Kathryn Mill
"Ultimately women with Turner's syndrome must take control of defining their identities. They must do the studies, write the articles, and make the speeches that tell the world who they are and what they can do'.

(Orten and Orten, 1994).
Background to the study

- Two part Ph.D study on the psycho – social aspects of women with Turner syndrome (part 1 subject group n = 113; part 2 subject group n = 8)

- Comparison group – sisters or friends of the subject group (comparison group n = 58)

- University of Tasmania, Hobart Australia 1992-93

- University of Sydney, Australia 1993-96

- University of Western Sydney, Australia 1997-2001
Aims of the study

- To compare any psycho-social differences between a group of women with Turner syndrome, and a comparison group of their sisters or friends.

- To elucidate the major difficulties experienced by women with Turner syndrome, and thus address those issues requiring attention.
Quantitative study (numbers)

PART ONE
Recruitment

Turner syndrome sample

- Turner Syndrome Association of Australia Ltd
- Turner syndrome support groups
- Letter to the NZ Medical Journal
- Article in the Sydney Morning Herald
- Cytogenetic Laboratories
Recruitment

Turner syndrome sample cont...

- Endocrinologists, Gynaecologists, General Practitioners
- Endocrine Clinic – Royal Alexandra Hospital for Children, Sydney
- Department of Endocrinology and Genetics, Adelaide Women and Children’s Hospital
- In Vitro Fertilisation Clinics, Sydney
- Coincidental Personal Contact
Recruitment
Comparison sample

- Sisters or friends of participant with Turner syndrome
Description

Turner sample

- 113 women with Turner syndrome
- Aged 18-56
- 88 Australian
Description
Turner sample

- 51 Participants from New South Wales
- 9 Participants from Tasmania
- 8 Participants from Queensland
- 11 Participants from Victoria
- 4 Participants from Australian Capital Territory
- 5 Participants from South Australia
- 25 Participants from New Zealand
Description
Comparison sample

- 58 women

- Sister or friend of respondent with Turner syndrome – matched as well as possible for demographic and social factors
Measures

- Turner Syndrome Questionnaire
- Demographics
- Health
- Career
- Childhood relationships
- Adult relationships
- Sexual arousal and behaviour
- Turner syndrome
- Coopersmith Self-Esteem Inventory
- Personal Attributes Questionnaire (PAQ)
Procedure

- Face to Face
  62 participants within the Sydney or Hobart region

- Self – Administered
  51 participants outer New South Wales, other Australian states, New Zealand
Areas of Investigation

- Demographic features
- Educational attainment
- Children/Employment
- Occupation/Work
- Career
- Physical health
- Mental health
- Childhood relationships
- Adult relationships
- Sexuality
Demographic features

- A significant difference occurred between the TS and the comparison groups with respect to whether they have children.
- Respondents from the Comparison group were likely to have more children than respondents from the TS group.
- A significant difference occurred as to whether participants were employed. The Comparison group were less likely to be employed.
- No significant difference in marital status, but of those who are married, a greater percentage of the comparison group report marrying at a younger age:
  - TS group (53% at less than 25 years)
  - Comparison group (78% at less than 25 years)
A greater percentage of the TS group than the Comparison group resided at home with their parents.

<table>
<thead>
<tr>
<th>Group</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>TS group</td>
<td>37%</td>
</tr>
<tr>
<td>Comparison group</td>
<td>17%</td>
</tr>
</tbody>
</table>
Educational Attainment

- Similar to sisters
  (Konradsen and Nielsen, 1993, Downey et al., 1991; Nielsen et al., 1977)

- Diligent attitude
  (Swillen, Fryns, Kleczkowska, Massa, Vanderschueren-Lodeweyckx, and Van Den Berghe 1993; Nielsen and Sillesen, 1981, Nielsen, Nyborg, and Dahl 1977)

- Attributed to determined attitude
  (Money and Mittenthal, 1970)
Educational Attainment cont…

- Similar to sisters
  
  (Konradsen and Nielsen, 1993, Downey et al., 1991; Nielsen et al., 1977)

- Undergoing higher education
  
  (Kerzner, 1998; Pavlidis, McCauley, Sybert, 1995; Sybert 1995; Aran, Galatzer, Kauli, Nagelberg, Robicsek and Laron, 1992; Orten and Orten, 1992)

- Achieved a higher level of academic education than similarly matched women from the general population or parents
  
  (Pavlidis et al., 1995; Sybert, 1995; Okada, 1994; Aran et al., 1992)
Lack of concentration

(McCauley, Ross, Kushner and Cutler 1995; Rovet and Ireland, 1994; Rovet, 1992)

Difficulty with mathematics and spatial tasks


Language ability level with peers

(Rovet and Ireland, 1994; Nielsen and Stradiot, 1987; Nielsen and Sillesen, 1981; Nielsen et al., 1977)
The majority of both groups report that they have not been encouraged into any particular occupations.

The T.S. group demonstrates a slightly lower, (but not significantly so) rate of encouragement than their comparisons.

Occupations encouraged to enter
The main occupation reported by the T.S. group is clerical/reception (17%) and the Comparison group is teaching (50%).

A greater percentage of responses from the Comparison group than the T.S. group indicate encouragement into nursing (21%) or teaching (50%).

A greater percentage of the T.S. group than the Comparison group were encouraged into child care (14%) and “other” occupations (41%) which included a varied, apparently non-systematic range of occupations: helping profession, arts, computers, sport, book-binding, librarian, business, science, medicine, dentistry, law and home economics.
Who encouraged into occupations

- Parents were reported by both the T.S. group (39%) and the Comparison group (44%) as most likely to encourage them

- The same percentage of responses from the T.S. group and the Comparison group indicate respondents being encouraged into occupations by their teacher/careers advisor (15%) and employers (4%)

- A similar percentage of responses from the T.S. group (4%) as the Comparison group (2%) indicate respondents were encouraged into their occupations by a vocational advisor or psychologist, and other relatives friends/acquaintances (T.S. = 17%; Comparison = 25%)
Reason encouraged into occupations

- Marked similarity was found between the groups with respect to being encouraged into certain occupations

- 4% of responses from both groups indicated that respondents were encouraged into occupations due to it being appropriate to their stature
Reason encouraged into occupations cont...

<table>
<thead>
<tr>
<th>Reason</th>
<th>TS No</th>
<th>TS %resp</th>
<th>Co No</th>
<th>Co % resp</th>
</tr>
</thead>
<tbody>
<tr>
<td>Interest and competency</td>
<td>31</td>
<td>32</td>
<td>20</td>
<td>39</td>
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<tr>
<td>Aptitude in a vocational assessment</td>
<td>9</td>
<td>9</td>
<td>4</td>
<td>8</td>
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<tr>
<td>Appropriate to stature</td>
<td>4</td>
<td>4</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Others did this work</td>
<td>7</td>
<td>7</td>
<td>6</td>
<td>12</td>
</tr>
<tr>
<td>Subjects best fitted</td>
<td>15</td>
<td>16</td>
<td>7</td>
<td>13</td>
</tr>
<tr>
<td>Competent in subjects</td>
<td>11</td>
<td>12</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Others suggested I apply</td>
<td>5</td>
<td>5</td>
<td>6</td>
<td>12</td>
</tr>
<tr>
<td>Considered all I could do</td>
<td>3</td>
<td>3</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Other</td>
<td>11</td>
<td>12</td>
<td>1</td>
<td>2</td>
</tr>
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</table>
Discouragement from occupations

- The majority of both groups report that they were not discouraged from particular occupations
Discouragement from occupations cont...

- Of those who were discouraged from entering a particular occupation(s), marked similarity in responses occurred between the two groups.

- The greatest difference occurred in the “other” category (T.S. = 34%; Comparison = 61).
Discouragement from occupations cont...

- Occupations Respondents Were Discouraged from Entering T.S. Group (n = 27); Comparison Group (n = 16)

<table>
<thead>
<tr>
<th>Reason</th>
<th>TS No</th>
<th>TS % resp</th>
<th>Comparison No</th>
<th>Comparison % resp</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aptitude in assessment</td>
<td>4</td>
<td>7</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>Not appropriate to stature</td>
<td>13</td>
<td>20</td>
<td>4</td>
<td>11</td>
</tr>
<tr>
<td>Considered unable to do the job</td>
<td>14</td>
<td>24</td>
<td>6</td>
<td>17</td>
</tr>
<tr>
<td>Not competent in subjects</td>
<td>6</td>
<td>10</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Considered could do better</td>
<td>5</td>
<td>8</td>
<td>8</td>
<td>23</td>
</tr>
<tr>
<td>Not appropriate qualifications</td>
<td>6</td>
<td>10</td>
<td>2</td>
<td>6</td>
</tr>
</tbody>
</table>
Of those who indicate that they were discouraged from a particular occupations, T.S. group (n=25) and the Comparison group (n=16), a similar percentage of responses from the T.S. group as the Comparison group were indicated, when respondents were asked by whom were they so discouraged.

Of those who indicate that they had been discouraged from a particular occupations, the main reason reported by the Comparison group (n=27) was that it was considered they could do better (23%). Only 8% of responses to this category were indicated by the T.S. group.
Discouragement from occupations cont...

- A similar percentage of responses occurred between both groups, with respect to all other categories
Working with children

A slightly higher percentage of respondents from the T.S. group (42%) than the Comparison group (34%) reported that they work with children.

Why work with children

- The main reason given by both groups (n=45; 60%) and the Comparison group (n=18; 73%) is that they enjoy working with children.
- The same percentage of responses in both groups (23%) indicate respondents work with children because they can identify with them.
- Eleven percent of responses from the T.S. group and four from the Comparison group.
Indicate respondents work with children due to their getting on better with children than adults

6% of responses from the T.S. group and none from the Comparison group indicate that respondents work with children as compensation for not having their own children.
Suitability to working with children

- No significant difference occurred with respect to suitability to working with children

- A greater percentage of the T.S. group (48%) than the comparison group (31%) consider themselves to be very suitable to work with children, and a smaller percentage of the T.S. group (4%) than the comparison group (11%) report they are neither suitable nor unsuitable to work with children
Work situation

- A similar percentage of responses from both groups indicate that respondents experience or have experienced difficulties:
  - relating to peers (25%; 21%)
  - relating to superiors (16%; 19%)
  - coping with the work (19%; 14%)
  - meeting the physical demands of the work (14%, 13%)
  - being taken seriously by others (20%; 22%)
  - difficulties relating to subordinates (6%; 11%)
Work situation cont...

- Respondents in the T.S. group are employed at a significantly higher rate than those in the comparison group.

- The T.S. group demonstrates a higher rate of employment than their comparisons 79% of the T.S. group compared with 70% of the comparison group is employed.
The occupation of respondents was coded according to Daniel’s (1983) scale. This scale categorises occupations on a scale of one to six, with one representing high prestige occupations, and six representing low prestige occupations.

The mean employment status was similar in both groups.
Career

- Career status
- A significant difference between the T.S. and Comparison groups, in terms of whether they have a career
- T.S. group 81% n = 89 Comparison group 69% n = 40

- Career interest
- T.S. group ‘extremely interested’ 66%
- or ‘quite interested’ in
- pursuing a career
- Comp group ‘extremely interested’ 71%
- or ‘quite interested’ in pursuing a career

- Specific career
- Little difference between the groups in terms of what type of career they have
Career cont…

Specific Career of Respondents T.S. Group (n = 89); Comparison Group (n = 40)

Specific career

T.S.Comparison       Nº       % resp   Nº       % resp
Nursing              17           18    9         21  
Teaching             10            11    5         12  
Clerical/receptionist 20            22     9         21  
Child care           8            9      1         2   
Public servant       2            2      0         0   
Other                36           38    18         44
With respect to such occupations as nursing teaching and clerical/reception type positions marked similarities occur between the groups.

A slightly higher percentage of responses in the T.S. group indicate child care work.
Reason respondents chose their career

The greatest percentage of responses from both groups indicate that respondents had chosen the career that was most suited to them.
A similar proportion of respondents in both groups indicate that they chose their particular career because they would have felt unfulfilled staying at home, that they were encouraged by a parent or teacher or that they couldn’t have children. Overall, marked similarity occurred between the two groups, with respect to the reasons for which they chose their career.
### Reason Respondents Chose Their Career cont...

**T.S. Group (n=77); Comparison Group (n=40)**

<table>
<thead>
<tr>
<th>Reason</th>
<th>T.S. Group</th>
<th></th>
<th>Comparison</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No</td>
<td>% Response</td>
<td>No</td>
<td>% Response</td>
</tr>
<tr>
<td>Suitability</td>
<td>54</td>
<td>37%</td>
<td>26</td>
<td>45%</td>
</tr>
<tr>
<td>Encouraged</td>
<td>18</td>
<td>12%</td>
<td>6</td>
<td>10%</td>
</tr>
<tr>
<td>Fulfilment</td>
<td>21</td>
<td>15%</td>
<td>9</td>
<td>15%</td>
</tr>
<tr>
<td>Childlessness</td>
<td>6</td>
<td>4%</td>
<td>1</td>
<td>2%</td>
</tr>
<tr>
<td>Financial</td>
<td>28</td>
<td>11%</td>
<td>10</td>
<td>17%</td>
</tr>
<tr>
<td>Other</td>
<td>18</td>
<td>13%</td>
<td>8</td>
<td>14%</td>
</tr>
</tbody>
</table>
Physical Health

- Significant difference between the groups in terms of suffering from physical problems
  - TS group
    - hearing loss/ear infections: 12%
    - osteoporosis: 6%
    - thyroid problems: 6%
    - high blood pressure: 5%
    - diabetes: 5%
    - heart problems: 6%
    - asthma: 3%
    - skin irritations: 3%
    - horseshoe kidney: 3%
    - osteoarthritis: 3%
    - other complaints: 1%
  - 36% responded
Physical Health cont...

- Comparison group: 14%
- Asthma: 38%
- Other complaints: 1%
Mental health

TS group

- mental illness: 6%
- depression/anxiety: n = 4
- anorexia/bulimia: n = 3
Counselling

- TS group: 34% responded
  - Depression: 19%
  - Relationships: 16%
  - Self-esteem: 17%
  - Grief counselling: 10%
  - Accepting/understanding TS: 5%

- Comparison group: 47%
  - Relationships: 41%
  - Self-esteem: 6%
  - Grief counselling: 3%
Childhood relationships

- Respondents from the Comparison group reported being teased at primary school and left out more at school than respondents from the TS group.
Adult relationships

- A similar percentage of both groups reported their relations with others as being good.

- A greater proportion of the Comparison group than the TS group described their relations with others as being excellent.

- Respondents in the TS group were less likely than respondents in the Comparison group to have ever had a boyfriend.

- Respondents in the TS group were less likely than respondents in the Comparison group to be currently in a romantic relationship.
More respondents from the TS group (71%) than the Comparison group (60%) reported being treated according to their age by parents.

A greater percentage of respondents from the Comparison group (20%) than the TS group (8%) reported being treated as younger than their age by their parents.

A greater percentage of responses from the Comparison group than the TS group indicated being treated according to their age by others:

- eg. teachers/lecturers TS = 63% Co = 83%
Adult relationships cont...

- Similar reporting of being overprotected occurred across the two groups
Adult relationships cont...

- A greater proportion of the TS group than the Comparison group reported having more female friends.

- A greater percentage of responses from the TS group (30%, n=22) than the Comparison group (23%, n=7) indicated that respondents spend more time with friends of a particular gender due to opportunity/availability.
Adult relationships cont...

- A greater proportion of the Comparison group than the TS group reported having more male friends.

- Of those who indicated that they had ever had a boyfriend, the Comparison group reported having their first boyfriend at a younger age than the TS group.
A greater percentage of respondents from the Comparison group (37%, n=21) began going out with males at an earlier age than respondents from the TS group (13%, n=15)
Sexuality

- Respondents in the Comparison group engaged in sexual activity more often than respondents in the TS group.
- Respondents in the Comparison group more frequently enjoyed having sex than respondents in the TS group.
Self-Esteem

- No significant difference occurred between the two groups in terms of self-esteem

- No difference occurred between the two groups in terms of their perceived masculinity or femininity
Information concerning Turner syndrome (specific to TS group)

- Majority of respondents (67%) were first told about having TS between 11 and 20, 14% were first told when they were over 21.

- Information received about TS:
  - A lot: 23 (20%)
  - Sufficient: 42 (37%)
  - A little: 39 (35%)
  - None: 9 (8%)
Information concerning Turner syndrome (specific to TS group) cont...

- Majority (64%) reported that they had received information about TS between the age of 11 and 20, 30% were over 21.

- Learned consequences of TS (ie. infertility):
  - 5 - 10: 6 (5%)
  - 11 - 15: 48 (43%)
  - 16 - 20: 41 (37%)
  - 21+: 17 (15%)
Hormone replacement therapy (specific to TS group)

- Half of respondents reported feelings of well being since commencing hormone replacement therapy

- Majority of respondents 93(83%) were still taking hormone replacement therapy

- 19 (17%) indicated they were not taking any hormone replacement therapy

- 11 discontinued due to side effects
  - 7 discontinued due to doctor’s advice
  - 3 discontinued due to already achieving puberty
  - 1 discontinued due to cost
Effects of Turner syndrome (specific to TS group)

- Relationships with others most frequently reported
- Infertility and physical development also frequently reported
- Majority (70, 62%) believed having TS makes it more difficult to enter a relationship with a member of the opposite sex
- (20, 18%) believed having TS does not make it more difficult to enter an intimate relationship
- (22, 20%) didn’t know whether having TS affected them entering romantic relationships or not
Effect of Turner syndrome (specific to TS group) cont…

- Majority (91, 81%) reported having TS had affected their life
- (21, 19%) indicated having TS had not affected their life

- Infertility impacted on life:
  - A great deal: 35 (31%)
  - In some respects: 58 (51%)
  - Not at all: 20 (18%)
Infertility

- Desire to have own children:
  - A lot: 38 (34%)
  - Quite a lot: 37 (33%)
  - Neither wish/not wish: 17 (16%)
  - Not particularly: 17 (16%)
  - Not at all: 2 (1%)

- The majority of respondents (41) believed their infertility meant they were more interested in children.
Qualitative study (words)

PART TWO
Background to the Participants

- Eight women with Turner syndrome
- All resided in Sydney, Australia
- Aged late 20s to mid 30s
- Two married
- Six single
Seven educated to tertiary level

All eight were employed

Five - helping profession

Three – business/admin

Five were studying part time

Seven were taking hormone replacement therapy
Procedure

- In-depth semi-structured interviews commencing with an open-ended question about the participants' experience of having Turner syndrome.

- Other questions or probes:
  - Tell me about when you were diagnosed.
  - Tell me about when you were a child.
  - What was school like for you?
  - What about work?
Procedure cont...

- What about your relationships with others?
- What about self-esteem?
- What about infertility?

- One to two hours duration
- Interviews were audio-taped
- Interviews were analysed using grounded theory
Areas of investigation

- Infertility
- Short-stature
- Romantic relationships
- Relationships at school
- Teasing
- Adult relationships
- Disclosure to others
- Medical treatment
Emotional impact of infertility

- **Group 1**  “The major bummer of the whole Turner syndrome thing” – most major impact – devastated The choice had been taken away from them

- **Group 2**  Sad but not such an earth shattering experience
Emotional impact of infertility cont...

- **Group 3**  Never wanted to have children anyway

- Expectations of motherhood taken away

- Role as a woman called into question
Information concerning infertility

- Denial/delay/limited information

- Finding out about infertility by themselves (ie. school text books, lessons)
Emotional reactions to infertility

- Grief
- Shame
- Emotional triggers (ie. crying at the sight of a baby)
Coming to terms with infertility

- Great difficulty
- Acceptance (i.e. people are a lot worse off)
- Reasoning (i.e. I am never going to have children)
- Religion (i.e. faith)
- Expression (i.e. crying, talking with a friend)
- Withdrawal (i.e. from the outside world)
Other means of acquiring children

- IVF (cost, procedure)
- Adoption
- Fostering
- Needs to be negotiated with husband or partner
Other people’s children

- Enjoy a relationship with other people’s children ("I do relate well to kids. Most kids respond to me, and I have fun when I am with them")

- Careers and occupations working with children ("tend to warm to me a bit more than they do other staff")
Short stature (being told to stand up, and you are already!)

- Significant challenge
- Only obvious feature of having TS
Disadvantages of short stature

- Being teased by others
- Being overlooked for service in shops
- Not being invited to pubs or clubs
- Needing to prove yourself
- Not being taken seriously
Disadvantages of short stature cont...

- Being discriminated against
- Difficulty with playing sport
- Easy to gain weight
- Difficulty reaching high objects
- Ill-fitting office furniture
- Pregnancy (ie. IVF)
Advantages of short stature

- Some sport (ie. gymnastics, jockey)
- Warming to children
- Looking younger
- Regarded as less threatening
Relationships at school

“I felt really different to the other kids”

- Teasing
- Effect on self-esteem
- Effect socially (withdrawal, loner, lack of social skills)
- Feeling different, like there is something missing
- Feeling not as good as others
Coping with teasing

- Ignoring
- Fighting back
- Making light of it
- Others get teased
- Relationships during puberty (secondary school)
Coping with teasing cont...

- Delayed maturation – (feeling different)
- Effect on self-esteem
- Changing rooms
- Boyfriends
- Ostracism and Rejection
Adult relationships

“I don’t want to be that unhappy kid any more”

- Easier
- More satisfied with social relationships now
- Greater opportunity to meet and interact with a range of people
Good working relationships
Romantic relationships

- Tend to happen later than other women
- Difficulty with developing the skills if you have never had one
- Partner may have had more relationship experience
Romantic relationships cont...

- Self-esteem – not good enough for a relationship
- More likely to be single
- Difficulty maintaining contact with married friends
- Infertile marriage – should I have got married?
Disclosure to Others

- Difficult – tended not to do it, only with close people
- Reluctant to disclose to people at work (ie. boss)
Medical treatment

“Heaven forbid a child be allowed to ask any questions”

- Diagnosis occurred at different ages but generally around puberty
- If diagnosis left later, delayed maturation – not developing like peers
- Follow-up visits to doctor – despised
- Would have appreciated follow-up counselling
- Visits to doctor mainly accompanied by mother
- Disempowerment
- Doctors made all the decisions
Medical treatment cont…

- Spoke mainly to the mother
- Visits boring and routine
- Student doctors present
- Would have preferred female doctors
- Medical information received from doctor unsatisfactory
- Intimidated by doctors
- Not informed about the Turner Syndrome Association from doctors
Conclusion

Study one

- Physical problems (especially hearing loss) greater in TS group than the Comparison group
- Greater difficulty with relationships especially intimate relationships in TS group than the Comparison group
- Other factors (ie. education, occupation, career) were relatively similar among the TS group than the Comparison group

Study two

- Infertility was the greatest factor affecting those with TS
- Other people’s children became important to those with TS
Conclusion cont…

- Short stature was a significant factor affecting those with TS
- Relationships were affected by having TS (ie. delayed pubertal development, infertility)

General

- People with TS live reasonably satisfying and productive lives
- Given the strength and determination inherent within their character they invariably will deliver