

## Authors

Margaret. R. Kyrkou,<sup>1</sup>  
Rebecca O'Rourke,<sup>2</sup>  
Joan P. Murphy,<sup>2</sup>  
Verity M. Bottroff,<sup>1</sup>  
Dennis S. Smith,<sup>1</sup>  
Hilary M. C. V. Hoey,<sup>2</sup>  
Roy Brown<sup>1,3</sup>

<sup>1</sup> Disability and  
Community Inclusion,  
School of Medicine,  
Flinders University,  
Adelaide, SA,  
Australia

<sup>2</sup> Department  
of Paediatrics,  
National Children's  
Hospital, Trinity College,  
University of Dublin,  
Dublin, Ireland

<sup>3</sup> University of  
New England,  
Armidale, NSW,  
Australia

## Correspondence

margaret.kyrkou@  
flinders.edu.au

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## Puberty, Menstruation and Pain in Australian and Irish Females with Down Syndrome

### Abstract

*Females with Down syndrome (DS) have generally been considered a homogeneous group, equated with females in the general population in terms of puberty, menstruation and ability to indicate pain. However, parents who related more closely to their females with DS, and often comparing with their sisters, were reporting otherwise, indicating that these aspects of development in females with DS are still poorly understood. The purpose of this study was to obtain information from parents of females with DS, and where they were agreeable, from the females with DS themselves, in order to be able to compare those findings against any published data. The researchers elected to use postal questionnaires, supplemented with face to face interviews in Ireland, aware that the information required was sensitive, and that such information tends not to be discussed openly. There were striking similarities and common themes generated by the 103 Australian and Irish parent respondents as well as 57 of their daughters with Down syndrome. Implications of the research relating to females with DS, and directions for future research are discussed.*

### Literature Review

#### Puberty

Researchers increasingly claim the influences on pubertal timing and rate of development to be multi-factorial and inter-related, the timing of onset and course influenced by 19 cascading genes (Topaloglu, Kotan, & Yuksel, 2010; Parent, Teilmann, Gerard, Juul, Skakkebaek, Topari, et al., 2003) but modified by environmental factors operating through the central nervous system. Other factors appearing to influence both age at onset and progression of pubertal development include nutritional state, general health, geographic location, exposure to light and psychological state (Grumbach & Styne, 2003). As well, low birth weight has been implicated (Van Wessenbruch & Delemarre-van de Waal, 2006).

Tanner (1962) described five stages of puberty, first the growth spurt, followed by breast development, pubic hair growth, menstruation, and finally axillary hair growth. However, in a book edited by Ash Monga (2006), ten respected gynaecologists pointed out that only 70% of females in the general population follow this pattern precisely.

Kaplowitz & Oberfield (1999) cited the Drug and Therapeutic and Executive Committees of the Lawson Wilkins Pediatric Endocrine Society recommendation that limits of normal puberty be set at 7–13 years for white girls, and 6–13 years for black girls. Garibaldi & Behrman (2004) considered breast development to occur at approximately 10–11 years, with growth of pubic hair 6–12 months later, whereas Styne (2007) implied that although the sequence of pubertal development is always the same, the age of onset and the time interval from one event to the next may vary.

Marshall & Ross (1977) stated that breasts and pubic hair do not usually develop simultaneously, and seldom progress at the same rate towards the adult state. They reported a study by Marshall & Tanner (1969) who concluded that the widely held view that breast development always precedes pubic hair growth in normal girls is erroneous. Pubic hair growth may begin before the beginning of breast growth in 15–17% of females (Joffe, 2006; Reiter, 1988), and was considered by Rosenfield (2002) to be due to lack of direct linkage between breast and hair development.

Pueschel (1991) and Schwab (1995) claimed the timing of puberty and the sequence of sexual maturation in females with DS to be comparable to that of females in the general population, with the exception of those with untreated hypothyroidism or chronic medical problems such as heart disease, which may result in delayed puberty.

### *Presence of Cyclical Symptoms Preceding Menarche*

The only noted references to mood disturbance preceding menarche (first menstrual period) were made by Saenger and Di Martino-Nardi (2001), and Styne (2007), with comments that some mood disturbance may be observed in females who develop pubic or axillary hair before eight years of age.

### *Time Span from First Sign of Puberty to Menarche*

Opinion varies on the interval between the onset of puberty and menarche. Marshall &

Ross (1977) claimed the average time interval between the first indication of breast development and menarche is about two and a half years, and that menarche rarely occurs less than a year after the breasts have begun to develop. Rosenfield (2002) claimed menarche may occur within months after breast development, but it is so unusual that its occurrence demands exclusion of an abnormal hyper-oestrogenic state.

Other researchers reported wide variations of time from the first sign of puberty to menarche: Irwin, Shafer, & Moscicki (2003) – 1.5–8 years, average four years, and Garibaldi & Behrman (2004) – 2–2.5 years, up to six years. Lobo (2007) and Rosenfield (2002) reported time from breast budding to menarche to be 2.3 years  $\pm$  1 year, but can range from 18 months to 5 years. Aranoff & Bell (2004) and Joffe (2006) stated menarche usually occurs within two years of onset of puberty.

It is of note that the earliest time span reported is 18 months, with all other researchers reporting time spans in excess of two years. Females with breast development first were reported to have menarche at 12.6 years, versus 13.1 years if hair development occurs first.

Rarely (Styne, 2007), a girl may begin to menstruate at an early age without showing other signs of oestrogen effect, one unproven theory being that the girl may have increased uterine sensitivity to oestrogen. Generally the period only lasts from one to six hours, after which the progression of puberty continues normally. Blondell, Foster, & Dave (1999) also referred to benign premature menarche as a rare and poorly understood disorder thought to be due to transient ovarian activity that is self-limited. Some of these cases were attributed to exposure to exogenous oestrogens of either a medical (such as oral contraceptives) or agricultural origin.

### *Weight Requirement for Menarche to Occur*

Opinions differ regarding the role of body weight in relation to puberty and menstruation. Frisch (2002), and Frisch & Revelle (1971) claimed the minimum body weight required for menarche to occur was 47 kg. It is now believed that body composition is more important than

total body weight in determining the time of onset of puberty and menstruation (Lobo 2007). Beckmann, Ling, Smith, Barzansky, Herbert & Laube (2006) and Grumbach & Styne (2003) considered the minimum weight to be between 38.6 and 48.2 kg, with a body fat proportion of 16–24% to sustain ovulatory cycles.

Grumbach & Styne (2003) stated that the age of menarche is earlier than average in girls with moderate obesity, compared with delayed menarche in girls who are severely malnourished. Lassek & Gaulin (2007), and Lin-Su, Vogiatzi, & New (2002), reported girls in the obese (more overweight) group to reach menarche at an average age of  $11.87 \pm 1.1$  years, and girls in the overweight group at  $12.14 \pm 0.9$  years, compared with  $12.20 \pm 1.3$  years in girls of normal weight.

Lassek & Gaulin (2007) claimed multivariate analyses have supported the alternative view, that skeletal growth is the main factor. However, they also showed menarche is more closely related to fat distribution than to total body fat, but that those reaching menarche despite low levels of total body fat have relatively more fat stored in the gluteo-femoral region.

## Menstruation

Menstruation occurs as the endometrium (lining of the uterus) is shed following failure of implantation or fertilization of the oocyte. It is the outward manifestation of the menstrual cycle, and results from the culmination of complex interactions and biofeedback mechanisms involving hormones and neurotransmitters mediated through the hypothalamo-pituitary-ovarian-genital axis. The first episode of menstruation is known as menarche.

Age of menarche in the general population is variably described by researchers as occurring between the ages of 10 years and 13.8 years (Pueschel, 1991; Biro, Huang, Daniels, & Lucky, 2008). Thomas, Renaude, Benefice, Meeüs, & Guegan, (2001) when reviewing studies published worldwide found the mean age of menarche to be  $13.53 \pm 0.98$  years. By comparison, females with DS are reported to reach menarche in the United States of America between nine and 15 years of age, average 12 years 6 months (Pueschel, 2006). In the United

Kingdom it is reported to be 11–12 years (Buckley & Sacks, 1987) and 12.5–13 years (Cunningham, 1996), whilst in Denmark Goldstein (1988) reported a mean of 13.6 years. Age of menarche is affected by race, total body fat, rural or urban living, climate and exposure to light, altitude, and birth order in large families.

## *Premenstrual Syndrome (PMS)*

PMS is defined as “The presence of recurrent symptoms before menstruation with the complete absence of symptoms after menstruation” (Dalton & Holton, 1994, p. 4). The more severe form, Premenstrual Dysphoric Disorder (PMDD, DSM-IV-TR 2000), is variably reported to affect 2–15% of females

Researchers and clinicians considered a high proportion of females in the reproductive age group experienced symptoms of PMS (Pernoll, 1994, 90%; Johnson, 2004, 40%; Borenstein, Dean, Yonkers & Endicott, 2007, 30.4%). However, they also questioned whether this was mostly physiological molimina (any symptoms other than bleeding that precede menstruation) rather than pathological PMS. They considered that more severe PMDD affected between two and 15% of females (Johnson, 2004, 3–5%; Llewellyn-Jones, 1989, 5–15%; Beckmann et al., 2006, 5%). Lentz, Woods, Heitkemper, Mitchell, Henker & Shaver, (2007) screened 72 females using daily symptom diaries for two to three cycles, and found 26 had low severity symptoms, 20 had PMS, and 26 had a premenstrual magnification pattern.

Richardson in a 1992 survey of 217 female university students aged 18–47 years found the five most common symptoms to be mood swings (76%), irritability (74%), lethargy (65%), tension (65%), and weight gain (61%). He also stated that although PMS is experienced by females in all cultures, the frequency and severity of symptoms varied between cultures. Lentz (2007) noted common findings in females with PMS to be a history of maternal PMS, suffering more stress, and not having borne a child.

Johnson (2004) considered the characteristic symptoms to be a mix of mood, physical and cognitive disturbances, but that the hallmark symptom was irritability. In contrast, Lee and Rittenhouse (1991) found that the only symptom

experienced in over 60% of nurses across all age groups was weight gain or swelling. Mason & Cunningham (2009) in a prospective diary study of females with DS found symptoms suggestive of PMS in 18–20% of the sample.

### *Pain in Females with DS*

Craig (p. 10, 2004) lamented ‘There remains unfortunate resistance to acknowledging pain in people who have difficulty verbally expressing their distress’. Hennequin, Morin, & Feine (2000) claimed people with DS are not insensitive to pain, but they do express pain or discomfort more slowly and less precisely than the general population, contrary to previous beliefs that they are insensitive to pain. This claim was supported by Henderson (1985) who claimed people with DS are slow and lack precision on timed tasks due to cognitive factors and delays at the neuromotor level, difficulties likely to be responsible for the delayed sensory threshold found in the study, and which could be mistaken for insensitivity. People with DS were considered able to localize painful stimuli, but made more errors in their avoiding movement response. Henderson did make the point that the individuals with DS were able to respond to cold pain, and indicate the general location of cold stimuli. Lind, Vuorenkoski, Rosberg, Paratanen, & Wasz-Hockert (1970) studied the vocal responses of infants with DS, finding they needed more stimulation to cause crying, and took longer to react. They found this lack of cry response increased with age, with children with DS over one year old showing none of the visual responses such as grimacing, limb movements, or breaks in respiration seen in normal infants. Job, Chambers, Craig, & Oberlander (2004) noted children with DS were delayed in acquiring the language used by typically developing children to describe painful distress.

### **Introduction to Current Study**

Parents, predominantly mothers, have frequently reported that their daughters with DS differed from sisters in regards to puberty, menstruation and indication of pain, claims not supported in the literature, yet parent responses revealed consistent themes.

In the current study responses to the questionnaires illustrate aspects of the menstrual cycle and its impact, from the perspectives of females with DS, and their parents. The detailed descriptions indicate close observation and knowledge of the female, and illustrate the challenges faced by parents, often without being able to access relevant professional advice. With much of the information recurrently echoed throughout the responses, overarching themes were extracted.

### **Method**

The study, conducted over a one year time span, encompassed both quantitative and qualitative research methods in order to maximise information gained – information not easily obtained in clinical settings. In Australia, information was sought from parents of females with DS by means of a postal questionnaire, also offering the parent the opportunity to request a self-report questionnaire for the female with DS to complete. Apart from the initial University Social and Behavioural Research Ethics Committee approval, the researcher was required to obtain specific approval from each agency, and each school willing to participate. In Australia, aware that agencies would not be able to provide names and addresses to the researcher for privacy reasons, the researcher first approached relevant disability and education services to ascertain whether or not they would be willing to add names and addresses to prepacked but unsealed packs, and include a letter of support for the study.

Conscious that potential respondents might find some of the questions very confronting if they received a questionnaire without prior notification, potential respondents in Australia were first sent a ‘Request to Participate’ by services, enabling respondents to consent to being involved by returning the signed consent form, and providing their contact details to be posted or emailed the questionnaire. The one page ‘Request to Participate’ had a reply-paid envelope with the researcher’s university address on it, and the researcher’s initials in the top corner so administration staff would not need to open the envelope in order to determine the intended recipient. This was particularly important as this study was conducted

through the Department of Disability Studies, and South Australia is a small enough state for many people with disability and their families to have met each other through mutual organizations and activities. The packs contained a letter from the principal Flinders University of South Australia supervisor, Dr. Verity Bottroff, and a one page letter from the researcher explaining the research.

The same questionnaire was used in Ireland, the only question changes being the addition of demographic questions regarding age of parents at conception, and the female's karyotype. The questionnaires were distributed through Down Syndrome Ireland, with relevant ethics approval, and potential respondents were informed through newsletters, and invited to participate. These females with DS also attended the Down Syndrome Medical Clinic, so they and their parents were already familiar with one of the researchers, which would have added to their confidence in participating. Researcher Rebecca O'Rourke, a final year medical student, also interviewed fifteen parents, usually the mother, and her daughter with DS, for her medical student elective. The principal researcher from Australia when visiting Europe was also offered the opportunity to meet with some of the females with DS and their mothers. She was able to meet six mothers with their daughters, and a seventh mother whose daughter declined to attend, saying she "didn't want to talk about that stuff."

## Results

Questionnaires were completed by parents or females with disability for 95 females with disability in Australia. The 95 females with disability included the 28 females with DS referred to in this paper, with 3 of those questionnaires completed by females with DS, the remainder by parents. In Ireland, 75 parents completed questionnaires, with 54 of their daughters with DS being identified as having reached menarche. Of those 54, 15 females with DS and their parents, predominantly mothers, were interviewed. The females ranged in age from 10 to 42 years of age (mean 20.4 years) at the time the Australian questionnaires were completed, and 11 to 33 years (mean 18.6 years) when the questionnaires and interviews were completed

in Ireland. In Australia, the researcher anticipated obtaining more exact details from parents of the younger females, but was pleasantly surprised to find many of the parents of the older females were also able to provide exact details

This research generated a wealth of both quantitative and qualitative data, which for ease of presentation is divided into headings, with both quantitative and qualitative findings grouped under the one heading for ease of reference. The major component of the qualitative data is presented as direct quotations from parents, as they vividly illustrate the significance of the quantitative data for them. Numerous comments implied the study was addressing an area of importance to the respondents, but at the same time many of the questions raised suggested respondents in Australia wanted timely information, but did not know where to access it, or even that it was available. Respondents in Ireland appeared to have more access to the information they wanted through the Down Syndrome Medical Clinic.

Although respondents could have just ticked the relevant boxes and made minimal comment, many of them spent considerable time providing those detailed comments. Qualitatively, a number of recurrent themes emerged: surprise and dismay when menarche occurred without observed signs of puberty, or in less than 12 months from the first sign of puberty; the impact of cyclical pre-menarcheal symptoms (symptoms reported approximately monthly until menarche); and the inability of some females with DS to verbally indicate that they were in pain when behavioural changes indicated they were in fact experiencing pain; mothers preparing their daughters for menstruation over a prolonged time span; variable support from education staff; and resorting to menstrual suppression when the female was unable to cope or get support outside the home.

### Parental Anxieties, and Preparation of the Female with Down Syndrome for Menarche

Not all respondents provided detail regarding preparation of the female with DS for menstruation. In Australia, eight mothers provided all preparation, supplemented by books and

videos. School staff supplemented information from mothers for another five females, whereas school staff appeared to have been the only source of information for one female. Additionally, six mothers accompanied their daughters to courses on managing menstruation offered by disability agencies, finding them to be very beneficial. In Ireland, the females with DS and their parents had ready access and support through the Down Syndrome Medical Clinic. Respondents made the following observations:

I believe the notion that women with a “dis-ability” are *SO* different than the general population is absurd! Disability is complex and degrees are immense and generalizing on any health issue can be dangerous.

If people realized and started giving information earlier (re periods, etc.) that would be helpful.

Books [do not emphasize] fertility and response.

Grandma used the term “curse.” Mother finds it difficult to communicate in a positive way about periods. A friend called it “her friend.”

Parents identified some very practical ways they developed to prepare their daughters for menarche and coping with ongoing menstruation, including planning well in advance of menarche. Ideas included preparing stories to explain menstruation, modelling the management of menstruation, practicing wearing a pad regularly long before menarche, and family and education staff breaking down the steps involved in the same way as for teaching any other skill. The following comments from parents graphically illustrate the time and effort some parents (predominantly mothers), and in some instances education and support staff have invested in preparing the female with DS for menarche, and helping her to manage menstruation:

Not all people with a disability have a lot of different things to us. My daughter copes well with her growing up and her period, it depends on how it is handled right from the start and before. I’ve spoken to her about periods from about eight years when she became curious about my period.

When she was 11–13 years we talked about it (menstruation) a bit. I explained that she had a little pocket in her tummy which collected food in case she had a baby one day. Each month the

food needed to be made fresh, and the old food thrown away. She had a baby hole in a very warm place between her legs, and this is where the old food was sent to be thrown away. The food might be all different colours but mostly it would be red like blood. She would need to wear a pad in her knickers so that the old food would not stain her clothes. When the day finally came she was completely unfazed by it, and went to school forgetting it was happening. I felt very pleased by the way it all went, but was shattered when I realized I had not taken enough time to teach her about what you do with a pad when you change it. This should have been such an important part of the process and I missed it out.

Two respondents referred to the benefit of either the mother or sisters, or both, allowing the female with DS to observe them managing their menstrual loss. As a couple of parents pointed out, this might seem gross to people unfamiliar with the learning difficulties of people with DS, especially those with intellectual disability (ID), but it is effective because it visually shows the female the processes and steps involved.

I made her aware of menstruation aged 8–9 years by modelling changing of sanitary pads. Whilst it might seem gross to normal people, it was an extremely effective way to teach my intellectually disabled daughter with Down syndrome about periods. When she began menstruating, there was no problem, as she just thought it was normal like mum.

[At age] 9–10 years I talked to her, looked at books, and took her with me when I had my period so she could see and know what to expect. She even practiced wearing a pad in the months leading up to getting her period. Her School Services Officer also talked with her and did practice runs re where to go, who to talk to, what to do. When the time came it was a very smooth transition.

Apart from the females in whom menarche occurred without preceding signs of pubertal development, one mother only began preparing her daughter with DS when she started her first period, even though she did not reach menarche until she was 15 years of age, and managed her own toileting:

Breast development, hair and menarche all occurred when she was 15 years old. Vomited on the day of menarche. Awareness aged 15 years

with menarche. Her mother then showed her how to change and dispose of her pad, and where to obtain a fresh one. She was independent for toileting.

The following description illustrates the extreme difficulties faced by parents when school staff did not understand that if the girl with DS was already showing early signs of puberty, it was most likely she would also reach menarche before her peers:

I provided puberty awareness when breast development was noted. I pushed for some school education in the special education environment, but was told this would not occur until my daughter was in years 5/6, and at that stage she was only in year 4. I would like to know more about period management at school, with carers and with siblings.

Staff in education settings varied from being extremely positive and supportive, to not wanting to have any involvement in either preparing the student for menarche, or providing assistance managing menstruation. As a consequence, parents reported their daughters, in consultation with their family physicians advising on the adjustment of pill packs, were either prescribed the contraceptive pill with the period planned to coincide with school holidays when parents would be able to assist the female, or even prescribed it continuously.

Periods very regular, but missed five days of schooling because couldn't get help managing pads or clothing.

Behind these brief descriptions from respondents lies a huge investment in planning, time, effort and commitment by parents, carers and other family members to make the transition from childhood to early adulthood as smooth and stress-free as possible for the female with DS.

### **Interval from Commencement of Preparing the Female with DS for Menarche, and Reaching Menarche**

Parents of the female with DS varied widely as to when they provided information about menarche and ongoing menstruation. Some planned carefully well in advance, to give

her adequate time to learn all the skills she would need without rushing or confusing her. It is understandable that parents of the younger females with DS showing the first signs of puberty aged eight and nine years of age had not attempted to provide information or prepare their daughters at that young age.

Even when the female with DS had good language, many parents began preparing their daughter up to five or six years prior to menarche, with many giving information at least two years prior to menarche to help ensure timely preparation if menarche occurred earlier than the two years traditionally advised.

### **First Sign of Puberty in the Girls with DS**

Mean age of puberty in the Australian sample was 11 years (range 8–15 years), which is not dissimilar to that in the Irish females with the mean age of puberty 11.2 years (range 8–15.5 years). Menarche without any preceding signs of breast development occurred in 11% of the Australian females with DS and 13% of Irish females. Only 48% of the Australian females with DS, and 43.5% of the Irish females followed the Tanner's pattern of pubertal changes with breast development with or without associated hair development as the first sign of puberty.

First sign of puberty was pubic hair aged 12 years, with menarche also aged 12 years.

First sign of puberty was her first period. Concerning periods, she is no different than ones without disabilities.

### **Short Time Frame from the First Sign of Puberty to Menarche**

From respondent comments, it was obvious that for some parents and their daughters, the short time frame, or even no time frame, from the first sign of puberty to menarche meant preparation was less than ideal. This is particularly important for females with DS when they mostly require a longer lead time to acquire all the skills they need to manage menstruation in a range of settings.

In the sample, where details were known, no females with DS reached menarche more than 24 months from the first sign of puberty, but 19% reached menarche within one month of the first sign of puberty, 38% reached menarche seven to 12 months after the first sign of puberty, and 33% reached menarche 19–24 months after the first sign of puberty was noted. More females whose first sign of puberty was breast development reached menarche in less than 12 months. Of interest were the twin peaks of time from the first sign of puberty to menarche, with one peak at 7–12 months, the other at 19–24 months.

Many clinicians appear to be advising parents of the females with DS based on what is commonly stated for females in the general population. The statistics above demonstrate that even if clinicians warned parents of females with DS at the first sign of puberty that menarche would occur in approximately two years, virtually all would have reached menarche by 24 months.

The impact of the very short time frame from the first sign of puberty to menarche is echoed in the following vignettes:

She wasn't aware of it because it takes me by surprise. I was ready for my older daughter, I explained, saw and read books when she began to have breasts, so she knew everything. But what happens, its (daughter's name) with a baby's body except for hair at pubic area and at 11 years old got her period, first before her older sister at 13 years old. I wasn't prepared or ready for her and even if she is 11 years old, her height is like an 8-year-old. I didn't think she would understand all these concepts and the meaning of it. So when it happened I explained to her that it was normal and that she was OK. She was very mature about that, no crying. She was listening to me and I explained how to put pads and how to take them out and put in the garbage. After that I phoned the Down Syndrome Association, and they sent me special information and a video to help me to explain it to (daughter's name). A lot of visuals, and very good.

She grew a bit of hair on the pubic area then literally two weeks later had her first period. It seemed all very sudden.

No signs of puberty, got her first period aged 9 years, breasts at age 10 years, after her periods.

At 10 years we were assessed by the endocrinologist that she wasn't reaching puberty. Three months later she got her first period, so it was a real shock. I explained it to her then.

No warning, periods started with severe pain and vomiting for up to four hours. She vomited anything she was given. Pain worse with chocolate.

First period - slap in the face - not expecting it, developing young. Not prepared for the "secret," the word secret was used so would not tell anyone.

### Cyclical Symptoms Preceding Menarche

More of the Irish females with DS were reported to have cyclical symptoms preceding menarche (87%) compared with 63.2% of the Australian sample. Recurrent physical symptoms in particular occurred for nine to 16 months preceding menarche in 70% of the Irish sample, but were only reported for 32.1% of the Australian sample. Considering there appears to be no mention of this in the literature for either females in the general population or females with disability, this was a surprise finding. However, with the cyclical persistence of these symptoms for many months preceding menarche, and the impact on the quality of life not only for the female with DS, but all those around her, it is a significant finding.

Respondents were asked about the presence of symptoms preceding menarche, allowing free comment rather than restricting choices to a checklist, in order to highlight the major issues from the parent perspective. Responses were predominantly behavioural, variably described as cyclical mood change; grumpy or cranky; more easily upset; less cooperative; behaviour change at a certain time of the month; weekly mood swings and headache for six months; mood swings and unexplained stomach pains; stomach cramps every month; moody, with a small weight gain, and boils in her groin area. From some descriptions, it appeared that the symptoms were an increase of the usual difficulties faced by females with DS, such as being uncooperative and stubborn, with depressive symptoms. One respondent made this comment regarding her daughter preceding menarche:

Preceding menarche interested in boys and makeup, in particular calling most men her "boyfriend." Giggling with her girlfriends (silly behaviour). More resistant at home regarding bed time routine and food preferences.

Cyclical physical symptoms included abdominal pain, vomiting, enuresis or incontinence. The following vignettes are examples provided by the respondents:

The only sign was mucus discharge in her undies.

Enuresis in months before menarche.

Menarche aged 12 years, but in the 12 months preceding menarche had evening vomiting every month or two, ceased once menstruation started.

During two months and a half before her period, she was tired. I could see that something was bothering her. Few weeks before, she told me her tummy hurts, told me more frequently in the two weeks before her first period, was quiet and grinding her teeth.

This description from one mother illustrates the need for treating clinicians to consider the role of puberty and menarche in relation to symptoms traditionally considered to have a medical or psychological basis:

She hasn't reached menarche yet, but for the past two years, around 20<sup>th</sup> of the month has exhibited cyclic symptoms. This was first noticed as diarrhoea (rarely experienced before and so it was surprising to be sick again in such a short space of time). Other symptoms have included being cranky, wetting her pants (having been toilet trained since she was 18 months) and cramps (when she has cramps she likes a warm face washer). The cranky behaviour was also noticed by the teachers as being out of character. One day was particularly bad and so I collected her from school and took her to the doctor who could find no medical cause for the symptoms. The doctor did do a blood test for hormonal changes but there were conflicting suggestions about when to have the test done - during the cranky phase or mid-cycle. The test was done mid-cycle and came back negative. It has been difficult to find out if I am imagining the cyclic symptoms. The doctors seem to think she was showing no signs of puberty and were therefore sceptical (she has since developed breasts to the extent of requiring

bras). Assuming the behaviours are related to periods, the difficulty then for me was what to do about that. I don't experience PMT myself and so had little knowledge. I am also reluctant to medicate unnecessarily and do not look to alternative medicines. When she broke her arm her pain response was very similar to the "cranky" cyclic response so I now believe that she has been experiencing pain. I think I was relieved when I felt I could manage this with pain relief rather than more complex hormonal treatments. Subsequent to completing the questionnaire, her mother informed the researcher that when her daughter reached menarche, her symptoms ceased.

Females in whom cyclical symptoms of vomiting or diarrhoea for the preceding year or so suggested they were related to menarche were reported to have lessening of their symptoms leading up to menarche, ceasing with menarche. Some of these females did report similar symptoms occurring when they were premenstrual.

### Weight of the Females with DS at Menarche

The following vignettes refer to females with DS whose weights were less than the minimum which Frisch & Revelle (1971), Frisch (2002) and Beckmann et al (2006) considered to be required before the female would reach menarche.

Where respondents were able to provide the exact weight of the female with DS, one weighed less than 35 kg, five weighed between 36 and 40 kg, and two weighed 41-45 kg, as illustrated by the following vignettes:

Underweight when she got her first period, and still underweight. Cycles mostly last 28 days, sometimes a little longer or shorter.

Weight about 38 kg when reached menarche, now 40 kg. Twin sister reached menarche one year earlier. Periods not regular, all over the place.

Was underweight at menarche, currently weighs 29 kg and has only had two slight periods in 18 months.

First sign of puberty was pubic hair and first period simultaneously at 12 years, weight was 35-36 kg.

## Menstruation

Mean age of menarche was 12.1 (10–15) years in Australia, 12.3 (9–15.5) years in Ireland, with 75% of females with DS in Australia, and 80% in Ireland reported to mostly menstruate regularly. In the sample of eleven females with DS for whom exact weights were reported, the only female reporting irregular cycles weighed less than 35 kg at menarche. Several respondents highlighted the fact that *their daughters were thrilled to reach menarche because it meant they were normal in at least one way*. All of the females with DS for whom a response was given were able to manage their menstruation independently. The following vignettes illustrate some of the issues females with DS, parents and support staff have to contend with:

Breasts at nine years of age. First period twice per month before she was 12 years old. Sometimes periods extremely heavy – sometimes two times in one month, now 26 day cycle, period for a good week.

First period at 11.5 to 12 years, had underarm and pubic hair before. Natural sister got her period at 15–16 years of age.

She said “If I don’t wear a pad, I won’t bleed.”

I hate them; I hate stupid periods; hates having her period; hates being uncomfortable and wearing pads.

When she did have periods, she would gag, etc. at the sight of blood on the pad.

Periods every four to six weeks, last five days, when has period spends extra time in toilet.

Terrible trouble with periods, stopped when she left school (stress) and lost weight.

Period comes more regularly and she doesn’t have facial hair when on a vitamin/mineral supplement called “Plus” (dioscorea and Ambrotose complex predominantly), but sometimes hard to get it into her.

Only three respondents specifically commented that their daughters coped well with menstruation, although a number of others implied that this was the case:

Has always been able to change her own pads and looks after herself quite well, and is hygienic (Comment from two respondents).

Periods initially regular, but at about 18 years of age developed irregular periods with considerable pain, later diagnosed as Polycystic Ovaries Syndrome. We were able to find the answers needed from specialists we selected-gynaecologist and endocrinologist:

When the female with DS reached menarche, her 7- [or] 8-year-old sister was devastated, even though she had been told about periods.

Mothers of females with DS nearing the end of their reproductive phase commented on a change in menstrual cycle patterns:

Periods initially irregular, became regular then reverted to being irregular.

Regular until about 40 years of age, then started to become irregular.

Mother would like to know about menopause training.

## *Premenstrual Symptoms Reported in Females with DS*

To avoid making the questionnaire too long and risking disengaging potential respondents, questions related to premenstrual symptoms provided the opportunity to tick boxes as well as add comments to amplify the information. This was intended to facilitate potential respondents reporting the symptoms which had most impact for them.

Due to the difficulty of distinguishing premenstrual symptoms resulting from Premenstrual Syndrome (PMS) from those caused by premenstrual pain, the term premenstrual symptoms is used rather than the more specific term of Premenstrual Syndrome. This was intended to overcome the difficulties females with DS have in being able to clearly articulate what they are experiencing, and because neither condition has clearly defined diagnostic features. Premenstrual symptoms were reported for 77.8% of the Australian sample, but only 56% of the Irish sample.

The following vignettes illustrate some of the difficulties faced by females with DS and their families during the premenstrual week:

Primary school staff noticed her behaviour deteriorated in the premenstrual week, becoming defiant and reluctant or straight out refusing to participate in work or play.

Skin erupts badly, clears up for about three days from beginning of her period. In the premenstrual week she withdraws socially (more than usual), and is more sensitive than usual. More reluctant to go to work, claims she gets tired because of her period.

Understandably many respondents reported the same issues premenstrually, so they have been grouped together when descriptions are similar. In order of frequency of occurrence, the most common symptoms reported by females with DS were fatigue; anxiety tension or irritability; mood swings; depression or crying; weight gain or swelling; and skin disorders:

Quite tired, bags under her eyes, sits around, can be a bit teary and cry for no reason, and then gets pain in her tummy a couple of days before her period starts.

Before period doesn't eat very much, just wants to sit. Acne, and face more purple before her period.

Tired, very tired in the week before her period, is sensitive and tears come easily.

Wants to rest more in the week before, and can get a little cranky before and during her period, just like the rest of us.

A bit more tired before her period, bad enough to miss four weeks study in a year. Mum called to collect her from school.

Tired for a couple of days before, not so cheerful, able to state she has pain.

Tiredness and depression. A bit teary in the week before, with low personal tolerance, but generally good pain tolerance; very sensitive, emotional, sometimes depressed, crying because she's sad.

Becomes a bit quiet and sleepy, goes to bed early maybe twice a week.

Occasionally tired and touchy, faint, dizzy, teary.

More irritable and stubborn. Sometimes stubborn on day before and tired, slightly moody and sulky.

In the week before her period wants to be with boys.

Ratty and loud, is upset easily, and stomach bloats up. Appears to have the same premenstrual behaviours as her sister: moody, becomes quite emotional and can get easily upset over things that normally would not affect her.

Quieter, lethargic, short-tempered and easily aroused, needs verbal prompting to keep on track of daily living skills, best to allow her more time; less tolerant before her period. Very moody, erratic, teary sometimes, pimples, very tired, complains of sore tummy.

Annoyed by the slightest of things, mood is terrible regardless of being ultra nice to her; more irritable the week before her period. Symptoms worse if has late nights. Pressing hard on the painful part of the body, was non-co-operative, cranky, irritable, unhappy, had difficulty concentrating or getting along with others, and was difficult to distract.

Puffy breasts, cries when period first arrives. May be feeling tense, but doesn't say so. Bloating, retains fluid, and is slightly moody.

Bloated stomach, with cranky irritable behaviour and tiredness which affects her quite considerably.

Every period vomited premenstrually and complained of nausea, problem dissipated with two Evening Primrose capsules following tea time. Evening Primrose all the time, omega 3. Suggested is good for PMS, and skin better.

It is interesting to note that compared with either the study by Lee & Rittenhouse (1991), or the one by Richardson (1992), for all symptoms reported in one or both studies, the frequency of those symptoms reported for the females with DS were markedly less than those for females in the general population. This may be partly explained by the fact that some symptoms rely on the female being able to both recognize and report symptoms such as breast pain, headache and backache in the absence of observ-

able signs. The only symptom not reported in females with DS was food cravings, but the rate of anxiety, tension or irritability, fatigue or lethargy, and depression or crying more closely approached the rates reported for the females without disability in the two studies.

Hot flushes were not reported in any of the females with DS, not even those thought to have reached menopause, but they were more frequently reported to exhibit mood swings, irritability or lethargy.

Factors reported to aggravate premenstrual symptoms in the females with DS in the study were stress, pressure and late nights, but not surprisingly more aggravating factors were described for females with DS who had good language. Of interest is the reported benefit of paracetamol in relieving premenstrual symptoms, suggesting the symptoms are pain related, but not necessarily recognized as such by the respondents. Paracetamol and other pain relievers were given to more of the females with good language skills.

Respondents identified a number of factors which relieved premenstrual symptoms in females with DS, mainly sympathy or comfort; heat bag; not being rushed; being left alone; and paracetamol:

I am unsure as I have never really taken notice. I wish I had. I feel I know very little about experiences of menstruation for women with disability. I can only relate it to my experiences and that of my daughters, and I am unsure as to whether that is appropriate or not. There is little information or education available. I am embarrassed about my lack of knowledge. All questions needed a "Don't know" option.

### ***Women with Disability on Menstrual Suppression***

Only 9% of Irish females, and 18.5% Australian females with DS had menstrual suppression, mostly with the oral contraceptive pill (OCP). Reasons given for use of the OCP were to either reduce the menstrual loss, or in one instance to prevent periods:

Contraceptive pill has definitely helped. She likes to be able to visually "see" when to expect her

period. Flow also a little lighter making it easier to manage.

She currently takes a birth control pill every day so she only has occasional break through.

Now on pill, goes off in the holidays to bleed for seven days.

Commenced low dose Provera in stage four of puberty. First period very light and short before Provera dose changed to suppress further periods. Her specialist (endocrinologist) has recommended that Provera be ceased and I intend to do this when she has finished meds (about six days away).

Depo Provera two weeks ago for first time; before Depo Provera emotional, teary, this time not getting early symptoms. Depo Provera great now, no pain, still gets periods when they are due. Now has to write when they are due as there are no symptoms.

### ***Frequency of Menstruation Related Pain***

Data were extracted from the large volume of information provided by respondents in relation to pain in females with DS. Whereas some females were able to describe their symptoms verbally, others were unable to do so, and had to rely on observant parents and carers being sensitive to minor changes indicating the presence of pain.

Twenty of the 27 Australian females with DS had menstruation related pain (74%), higher than that reported for females with other disabilities in the study, and higher than many females in the general population, in whom the rate of period pain (dysmenorrhoea) is reported to range from 53–85%. Irish females with DS reported an even higher rate of 80%. Surprisingly, although 85.7% of the females with DS were able to state they had pain when hurt or unwell, only 35.7% of those were also able to state they had pain premenstrually or in relation to menstruation. These results indicate that to assume that a female with disability who has good language skills will consistently be able to state she has pain under all circumstances is erroneous. The following scenarios illustrate the impact of menstruation related pain on the female with DS:

Has missed about 15 days of school due to pain; has missed five to six days of school in past year, but stomach pains on the first day of each period if at school would be enough to send her home; misses one week every month when period starts on a school day; pain with most periods, has missed about seven days of work because of pain.

I would love to find something to help S (apart from the contraceptive pill which she strongly refuses). I feel it is the severe contractions when the period starts that cause her to vomit. Sometimes has a bowel clearout as well. The difficulty is compounded by her irregular cycle. During the first 18–24 months of having her period there was no pain or vomiting.

No pain for years, then put on weight and the pain increased. When she lost weight the pain decreased, she was just moody.

Doubles up with period pain. Can be moody. Has no sim card in her mobile because of her visual impairment. Care workers tell her to go into another room, and “ring” her mum and they listen into the conversation to find out what the problem is.

The following vignette illustrates the practical difficulties encountered when a female with DS has good language, and it is assumed erroneously that she will be able to state she has pain.

Yells, kicks, screams (for severe pain only-broken arm). Six months ago when she broke her arm her pain response was very similar to the “cranky” cyclic response so I now believe that she has been experiencing pain. Now if there are symptoms, I treat her for pain. Surprisingly to me, as she is a very articulate person, she was unable to say her arm was hurting. I still feel really bad about this. I have never broken a bone and had no idea how much pain she was in. It took hours to get the morphine she needed. Still I now know that she is unable to tell me (or I need to teach her to understand pain signals) when she is in pain.

Face pale with pain.

Never complains of pain, has to be asked even if severe pain. More pale or washed out looking.

Pain inside and in hips-uses disprin, hot water bottle, cup of tea.

Severe cramps until after she left school. Gynaecologist consult with little success.

### *Behaviours Considered to Indicate the Female with Disability Was Experiencing Pain*

Respondents completed the Non-communicating Children’s Pain Checklist-Revised (Breau, McGrath, Camfield, & Finley, 2002) modified with the substitution of “female” for “child.” The checklist included both verbal and non-verbal indicators of pain. Respondents reported 92.5% of Australian females and 85% of Irish females with DS exhibited a number of behaviours thought to indicate that the female was in pain.

The ability to point to or touch the part of the body that hurt for any reason in females with DS showed 60.7% were able to point to pain when hurt or unwell, but only 17.9% were able to point to premenstrual or menstrual pain. Only 10.7% of females with DS were reported to bend forward when in pain for any reason, but from clinical experience of observing typically developing young females with period pain, the researcher would have expected a higher percentage of females to bend forward with pain. Although 17.9% were reported to press on the painful area when hurt or unwell, only one female pressed on her abdomen when premenstrual. Other features observed when the female with DS was thought to be in pain were absence of smiling (25%), mouth turning down (25%), or teeth grinding (10.7%).

More females with DS were reported to be non-co-operative, cranky, irritable or unhappy, jumping around, agitated or fidgety when premenstrual or menstruating than when hurt or unwell, the opposite of most behaviours reported for other females with disability in the larger Australian study. Three females with DS screamed or yelled very loud, lips were puckered up tight pouting or quivering, brow was furrowed, and they were floppy, stiff, spastic, tense or rigid when in pain or unwell, but not when premenstrual or during menstruation. Two females were reported to have facial pallor when premenstrual or menstruating, which suggests they were in pain.

### *Factors Aggravating Period Pain*

Respondents reported a number of factors which they considered to aggravate period pain, namely excess movement, stress or pressure, late nights, using a heat pack or the sight of blood. Not surprisingly, factors aggravating period pain were more easily identified in females with DS who had good language. The following vignettes illustrate difficulties faced by females with DS and their families when the female has pain.:

I find she complains more and seeks support (i.e., medication, bandage, and emotional support) than a normal child would.

Pain aggravated by sitting and standing, eased by heat bag, rest in bed, Panadeine or Ibuprofen with codeine.

Tried Ponstan and Naprogesic which made her sleepier and more nauseated. Missed at least one day of school, sometimes two days, every cycle due to intense period pain with nausea.

Pain aggravated by having to remain active, only wants to lie down and sleep. Because she hasn't learnt to take tablets I can only give her soluble paracetamol at present, whereas actual period tablets would be better.

Moans and cries with general pain.

### *Factors Relieving Period Pain*

Respondents were also asked about factors relieving period pain, citing rest, sitting quietly, using a heat bag or hot water bottle, distraction, alternative treatments, sympathy, comfort, and being left alone as being helpful. Paracetamol, Ponstan or other period tablets were used by 60% of the Australian females, and 70% of the Irish females. Of interest is the 33.3% of females with DS and good language given pain relief compared with only 14.8% of those with either poor expressive language, poor receptive language, or both.

### *Reactions to Pain*

Two respondents reported a paradoxical response to pain, appearing more to react to realizing the presence of injury rather than actually feeling pain.

She has a low pain threshold and resists medical treatment-if minor cut or abrasion, seeks help and comfort straight away.

In reaction to general injury is stoical, will sometimes indicate pain, and sometimes small injuries elicit a large response.

This female with DS was noisy when in pain, unlike many of the females with DS, who were more likely to be quiet and not wanting to move around much.

With pain of general injury cries or yells depending on how much it hurts, or the severity of the pain.

Other respondent reports illustrate the impact of pain on the female with DS:

Gets over pain faster than siblings.

Copes very well with general injury, doesn't react as much as a person without a disability would. Does get scared though. Reacts more to be scared than actual pain.

Hard to tell when she has pain, she seems to put up with pain for several days before casually mentioning that she has pain. If it is sharp pain in the night she will say straight away.

The respondents providing the following quotations do not seem to be aware the female has pain, but the fact that she goes quiet with general injury, or is quiet and withdrawn for the first two to three days of her period implies that she is in fact feeling pain. Only accepting pain killers once or twice is more likely the result of the individual not understanding that she has pain, and that the painkillers will ease it for a while. It could also be that when she is feeling low, she does not want to be bothered by being pressured to take tablets.

With general injury goes quiet. I am not sure how much pain she feels-her symptoms are more emotional, and ease with Nurofen and sympathy.

She is quiet and withdrawn the first two to three days of her period, but usually only accepts pain-killers once or twice. Recently hasn't wanted to go to school on the first day.

### *Reported High Pain Threshold*

High pain tolerance was reported by 11 respondents and minimal reaction to pain was highlighted by four respondents. However, the researcher gained the impression that it may be those females with DS, not able to make others aware of their pain, when from respondent reports they are showing definite behavioural indicators of pain.

Respondents considering the female with DS to have a high pain threshold may reflect the fact that the presence of pain is not recognized if the female does not state she is in pain. Many of those females are obviously suffering, and it is important for all those relating to the female with DS to pick up on the non-verbal indicators, and support the female appropriately.

Pain with most periods, but very pain tolerant, it has to be pretty bad to complain; pain with most periods, but very little reaction to pain, able to state she has pain, and point.

Has a very high pain threshold, but able to state she has pain. Only has occasional pain with periods, but eased by comforting and heat bag.

She takes a lot before she will complain of pain. During her period her face was pale, doesn't feel pain as much, and never appears to be in pain.

Has lots of ear infections but I don't know about it until there is goo coming out, but will tell me if she has a headache. Above normal pain tolerance, ear infection only recognized when examined by doctor.

Rarely says she has pain, doesn't bend over, but moans whines or whimpers, eats less, not interested in food, doesn't smile, is less active and is quiet, more irritable and stubborn. When has inflamed gums no complaints of pain.

Says she has pain in the week before her period, points to her abdomen before her period, whimpers and seeks comfort when unwell.

Tolerates pain maybe a little bit better, able to state she has pain, and point.

Able to state she has pain and point, but doesn't usually complain much. Excess movement seems to aggravate period pain, rest eases it.

### **Discussion**

The present study contains examples of innovative ways parents have devised to assist their daughters with DS to cope with this often challenging phase of life, and has the potential to assist parents prepare their younger females with DS for puberty and menstruation. It also heightens sensitivity to the non-verbal indicators of pain, and indicates that to assume that a female with DS who has good expressive and receptive language will be able to realize she is feeling pain, and will state this, may not be true.

In Australia a postal questionnaire was considered to be the most appropriate means of collecting the information, as parents may not have had previous contact with the researcher, and be concerned as to how the sensitive responses might be used. It is interesting to note that although 71.4% of the females with DS in the present Australian sample were said to have good expressive and receptive language skills, and parents were given the opportunity to also request a questionnaire for completion by the female with DS, only three females with DS completed a self-report questionnaire. By comparison, in Ireland, the females with DS regularly attended the Down Syndrome Medical Clinic, with the opportunity for timely information provision leading up to puberty. They would also have known one of the researchers, increasing confidence in the study. Additionally, some of the details requested in the questionnaire were likely to have been recorded in the clinical records of the females with DS. The face to face interview also allowed for clarification of responses.

Although respondents were more likely to be the more literate or motivated parents, there is no obvious reason why the findings are not able to be cautiously generalized. These data provide an important social comment on the extremes involved in parental awareness and expertise ranging from those in desperate need of knowledge and support, to those well able, when heard, to advance professional knowledge. It may well have only just begun to tap the sensitivity, creativity and wisdom of a larger body of sophisticated parents, and similarly may well have only begun to document the frustrations of those still left in the dark.

Feedback from respondents has indicated parents welcome this research on a topic little known, but even less talked about. The overall impression is of parents wanting to help the females with DS, where capable of doing so, to cope with puberty and menstruation without traumatising them. It is important not only for parents, carers and staff to be aware that the female with DS may exhibit cyclical mood changes or physical symptoms preceding menarche, but also treating practitioners, in order to avoid the distress of inappropriate investigations and treatment. Although some parents and their daughters accessed some information and training opportunities in Australia, the ones named were metropolitan based and therefore not accessible for families living remote to the metropolitan area.

The researcher also gained the impression parents in Australia appeared to be working in isolation when planning how to support their daughters. Whilst parents generally understand their daughters' abilities and preferred modes of learning in a situation like this better than outsiders, they seemed to be planning individually without the benefit of any written information to work from. The question then becomes, was the information available not relevant to their situations, or did they not know where they could access such information. Parents referring to videos mentioned one of two videos, one called "Janet's got her period," and one developed specifically for females with DS. Janet's got her period is an old video which was developed for girls in the general population, and does not cater to females with DS who generally have less ability to either comprehend or generalize, as the language level would be geared at upper primary students able to think in the abstract. The video for females with DS was developed specifically for their learning needs and style, but parents might not in all instances be aware of its existence. By comparison, parents and their daughters regularly accessing the Down Syndrome Medical Clinic in Ireland should be able to access timely information and resources.

All who relate to the female with DS need to be aware that apparently good language may not mean that the female understands when she is in pain, and will be able to communicate that effectively. She may not even be aware that she

needs to make adults around her aware she has pain so she can be offered appropriate relief. It is distressing to consider how many females with DS who were in fact in pain were considered to have a behaviour problem, and managed with behaviour modification techniques. This would only serve to increase the female's distress.

### **Limitations of the Study and Recommendations for Future Research**

The study sample was not random, and relied on parent perceptions and memory recall. With the exception of the three females with DS in Australia who completed self-report questionnaires, respondents were providing proxy reports. Although 71.4% of the females with DS in Australia were reported to have good expressive and receptive language, it is unknown whether parents of the other 25 females with disability offered their daughters the opportunity to complete a questionnaire and the female declined, or the parents felt too embarrassed to raise the subject with their daughters. Whereas typically developing females tend to pick up information from peers, or from reading magazines, those with DS are less likely to. Professionals should be cognizant of this sensitivity, and provide information in ways parents of females with DS, and the females themselves, are able to access and understand.

This study was exploratory, to investigate whether informal comments from parents of females with DS represented the difficulties and challenges for only a few females with DS, or a larger cohort. The consistency of responses from two continents would seem to suggest the latter. As these are such important issues for the females with DS and all who relate to her, with no references in the literature alluding to the differences compared with typically developing females, a prospective study is required to more comprehensively explore these issues.

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Last but not least, very grateful thanks to my husband Steve, and my daughters Catherine and Nicole, for encouraging and supporting me by taking on household tasks. Catherine has autism and ID, and is not able to communicate very much, and it took me a while to pick up on the subtle non-verbal indicators that indicated she was in pain, even though I have medical qualifications. Consequently it is imperative to get this information out to parents and their daughters with disability, to minimize their suffering.

## Key Messages From This Article

**People with disabilities:** Your body changes as you grow up, as for all females. You need to be given the same information as for other females, but you might need more time to learn it.

**Professionals:** Females with DS and their parents have unique needs for understanding and information provision approaching puberty. Even those females with good language may not indicate pain in expected ways, so nonverbal indicators need to be considered. The old paediatric axiom "Listen to the mother" is very relevant in this situation.

**Policymakers:** In general, females are reaching menarche earlier than previous generations, and the same applies to females with DS. Education needs to keep pace with biological changes.

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