BACKGROUND TO THE SURVEY

The Child Growth Foundation provides a support group to parents and children with a wide range of growth related conditions. One of these conditions is Precocious Puberty among its variants.

75 members have been involved with the CGF with these PSM conditions, and the opportunity was therefore taken to build on the ‘anecdotal’ stories which surface every year at the CGF Annual Conference, to achieve two aspects:

- Advise the Medical Profession of Current Perceived Practice
- Educate its Members.

COVERAGE OF THE SURVEY

The Survey was designed to elicit opinions and information in the following areas:

- Initial Diagnosis and GP Practice
- Specialist Referral
- Treatment Options and their Management
- Impact at School
- Social Implications of the Condition.

In each of these Sections, respondents were asked to identify their experiences, evaluate their practitioner, and identify good practice.

RESPONDENTS

The Survey was circulated in April 2000, and 30 replies were received. The results are based on this sample.

GENDER

Of the children, 26 were female, 4 male. This reflects the greater prevalence of this condition in girls, and is indicative of the very serious nature of it in boys.
**AGE RANGE**

The current age range is 2 years to 26 years! All the children first demonstrated signs of PSM before age 8 and 9 respectively for girls and boys. Half are still under these ages.

**RANGE OF CONDITIONS**

Of the 30 children –

- 23 have Central Precocious Puberty
- 1 child has Premature Adrenarche
- 1 child has Isolated Premature Menarche
- 1 child has McCune Albrights Syndrome
- 4 Children have Hamartoma (Brain Tumours).

All the boys have Hamartoma, and this confirms the pathological association of this condition in boys, and the spontaneous non-pathological derivation for many of the girls.
FIRST PHYSICAL SYMPTOMS AND AGE OF SYMPTOMS

For GIRLS, three symptoms were typically demonstrated. These were:

- Pubic hair
- Breast development
- Bleeding or menstruation

For BOYS –

- Testicular development and pubic hair was demonstrated.

Associated with these “obvious signs” were other characteristics such as

- Sweating and body odour
- Underarm hair
- Discharges and urinary infections (girls)
  Height and general body size/development
- Mood swings (girls) and aggression (boys).

Some of these symptoms were demonstrated at birth for some of the children, and were initially shown from 6 months to 7 years for the others.

PARENTS RESPONSE AND TIMINGS

The GP is the typical “port of call” for the parents, and they seek the doctor’s opinion immediately on noticing these characteristics. Obviously not all the children demonstrated all of these characteristics on initial visits, but 2 or 3 of the symptoms will have shown themselves.

School doctors/nurses occasionally notice early symptoms (3 out of 30), and refer the parents to the GP.

GP’S RESPONSE AND EXAMINATION

Two-thirds of children were examined visually, but referral to growth charts was not evident at this stage. The other one-third were not initially examined by the GP.

The GP does not generally indicate a diagnosis, but may refer the matter to the next level or seek to reassure the parents. Delays range from 1 month to 2 years! The main difficulty is gaining acceptance of the concerns:-
“I will refer her for tests, but nothing can be done, but look out for bullying”
“Don’t be concerned” (the doctor saw us as pushy and demanding)
“Showed no concern and no apparent interest in finding out”
“The problem is very serious – we were frightened by the lack of information”
“Offered no advice and seemed unsure what to do next”
“He thought my daughter may have taken my pills”
“The bleeding is in your mind”
“Go away and forget about it – you’re concerned about nothing”.

However, good practice was evident in a number of cases (30%), and this was demonstrated by:

- Visual examination and discussion
- Weight and height taken and reviewed
- Good listening and concern demonstrated
- Sympathetic
- Explanation that referral was appropriate
- Advice that children grow at different rates

REFERRAL TO SPECIALIST

Typically this will take 1 to 6 months to organise dependent upon location/access to specialists etc, and the severity of the symptoms demonstrated, eg menstruation accelerates the referral/diagnosis.

The assertiveness of parents had an influence, particularly where the parents managed to acquire information from the Internet, the Child Growth Foundation, or other sources. Quotes include

- “Had a long battle (18 months) to gain referral
- “Had to demand a referral”
- “No referral was offered until 3 years”
- “Referral after menstruation”

This underlines the key role that the diagnostic phase plays in this condition and delays do not tend to occur after referral, but leading to referral.

DOCTORS RATINGS AND GENERAL COMMENTS

Respondents marked their doctors’ diagnostic processes etc. as:

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<tr>
<th>Rating</th>
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<th>%</th>
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<tbody>
<tr>
<td>VERY GOOD</td>
<td>33</td>
<td></td>
</tr>
<tr>
<td>GOOD</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>ADEQUATE</td>
<td>30</td>
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<tr>
<td>POOR</td>
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<tr>
<td>EXTREMELY POOR</td>
<td>17</td>
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</tbody>
</table>
On the **positive** side, the responses included:

“I feel we have a very good GP who is always cautious particularly with children”

“If it wasn’t for my GP, I don’t know what might have happened”

“Although my GP did her best it took us two and a half years to diagnose her condition”

“The overall handling of the situation was not very helpful, but I do believe they didn’t know what they were doing”.

On the **negative** side, the responses included:

“I have been made to feel that I am telling lies and exaggerating issues”

“Eventual referral was marked ‘routine’, causing additional delays”

“For two and a half years we were told that our son was tall because we were both tall, and we (the parents) could see a psychologist if we couldn’t cope with our son’s mood swings (no diagnostic tests were done for 2 and a half years)”

“I have not been back to see my GP, and I don’t think I ever will”.

Perhaps the most telling quote was from one parent who said

“We were very frightened as we didn’t know what was happening and why this was happening”

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**KEY LEARNING POINT:**

DO NOT DISREGARD DIAGNOSTIC SIGNALS AND LISTEN TO PARENTS. DO NOT BELITTLE CONCERNS OR THE CONDITION

**KEY QUESTION:**

HOW CAN THE PROFESSION SPEED UP THE REFERRAL FROM THESE OBVIOUS DIAGNOSTIC SYMPTOMS?
SECTION 2 - SPECIALIST REFERRAL

REFERRAL PATTERNS

26 of the 30 respondents were referred for specialist treatment, confirming the serious nature of the condition. Many of these were initially referred to General Paediatricians (25 out of 26), but the majority (19 out of 26) was subsequently referred to Paediatric Endocrinologists.

CONFIRMATION TESTS

The most frequent tests referred to were

- Blood tests (LHRH tests)
- General examination and height/weight assessment
- Left wrist bone x-ray
  General examination.

Pelvic ultrasound for girls was also a common test, and half the respondents recall MRI/brain scans.

DISCUSSION OF RESULTS AND LONG TERM ISSUES

About one-third of respondents did not feel that results were discussed, with two-thirds recalling limited discussion.

Examples of “good practice” include:

  “Further treatment will depend upon progression of bone age”
  “Consultant explained the pros and cons of alternative options”
  “Choices were explained – with or without growth hormone”

Examples of “poor practice” include

  “No treatment is available – your daughter will have a long and hard adolescence”
  “Nothing can be discussed with you”
  “No – it is too far down the road”
  “It took 2 years to confirm a diagnosis”.

Three parents were advised that there might be a possible reduction in their child’s fertility/long term fertility problems.

SECOND OPINIONS

Most children seemed to move from General Paediatrician to Specialist referral fairly quickly, although some parents had to revert to Private Care to
progress this route. Half the respondents however sought the need for second opinions.

**ADDITIONAL SOURCES OF SUPPORT/ADVICE**

The Child Growth Foundation and the Internet are the two primary sources of additional support/advice. The Magic Growth Foundation in America was also mentioned (x3) and the advantage that the CGF brings with it access to other parents was specified (x5).

In two cases parents were offered counselling/psychologist appointment on an active basis.

**SPECIALIST RATINGS AND COMMENTS**

Respondents marked their Specialists as follows:

<table>
<thead>
<tr>
<th>Paediatrician</th>
<th>Paediatric Endocrinologist</th>
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<tbody>
<tr>
<td><strong>VEN GOOD</strong></td>
<td>25%</td>
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<tr>
<td>GOOD</td>
<td>21%</td>
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<tr>
<td>ADEQUATE</td>
<td>25%</td>
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<tr>
<td>POOR</td>
<td>16.5%</td>
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<tr>
<td>EXTREMELY POOR</td>
<td>12.5%</td>
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<tr>
<td><strong>TOTAL</strong></td>
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The trend was for the Paediatric Endocrinologists to receive more favourable and reliably favourable ratings, particularly when referral patterns move from the Paediatrician to the Endocrinologist.

On the positive side for Paediatricians, responses included:

"The Paediatrician has developed a good relationship with our daughter – which is now building up with the Endocrinologist"

"The Paediatrician is extremely forthcoming with information"

"Our Paediatrician was honest and sympathetic he was not an expert in the area, but would refer us"

"The immediate tests that were necessary were done and organised straightaway"

"She is now treated with respect as she has grown older – they ask her how she is feeling!"
On the negative side for Paediatricians, responses included:

“We were thought of as fussy parents, but after scans for our son were immediately referred to Great Ormond Street to an Endocrinologist”

“Being told no treatment was available was devastating (child 9 and a half, who showed first symptoms at 6)”

“The Paediatrician was quite blasé – he offered no alternatives”

“The MRI scan was only offered on our insistence (to boy with hamartoma)”

The obvious inference is that the conditions are sufficiently rare that Paediatricians are not in the best position to confirm the diagnosis or distinguish pathological/non pathological variants, unless there is particular knowledge.

On the positive side for Paediatric Endocrinologists, responses included:

“The P.E was wonderful, understanding, diagnosed our child’s condition”

“The Endocrinologists have all been excellent we trust them, and they don’t talk down to us”

“If an Endocrinologist had been consulted, perhaps more could have been done”.

On the negative side for Paediatric Endocrinologists, responses included:

“The specialist talks to us, above our child’s head, and is not very sympathetic to her”

“The Endocrinologist was dismissive and unhelpful – we sought a private appointment in London”

If I start to get angry , things get sorted out!
KEY LEARNING POINT

GET REFERRAL TO AN EFFECTIVE PAEDIATRIC ENDOCRINOLOGIST AS SOON AS PRACTICABLE - ROLE OF PAEDIATRICIAN IS TO RULE OUT PATHOLOGICAL CONDITIONS BEFORE EXPERT DIAGNOSIS

KEY QUESTION

HOW CAN THE PAEDIATRICIANS AND PAEDIATRIC ENDOCRINOLOGISTS DIFFERENTIATE THEIR ROLES IN REFERRAL – OR SHOULD WE BYPASS THE PAEDIATRICIAN?
TREATMENT OPTIONS

23 of the children opted for treatment. For the others, they either chose not to take treatment, or non was available.

22 of the 23 children opted for Zoladex/Prostrap Injections, with an additional child also taking daily growth hormone. Other associated actions include organic foods, dieting and evening primrose oil to combat polycystic ovarian syndrome in later life.

START OF TREATMENT

From 13 months to 9 years. There are two peaks at 3 and a half and 7 years. Most receive a 4 weekly injection (80%); some a 3 weekly injection (17%), and one every 12 weeks (3%).

CONTROL OF PUBERTY

In some cases it was considered too soon to tell, but in the majority of cases the following improvements were identified:

- Menstruation stopped
- Rate of growth stabilised
- Skin and greasy hair improved
- Fewer mood swings.

The stabilisation of growth was most marked for those children with hamartoma.

Emotional behaviour and anxiety appears to have reduced with the treatment, and sleep patterns improved.

ONGOING MONITORING ARRANGEMENTS

Most children receive the injections at the local surgery, but some from hospital where the relationship with the Endocrinologist’s nurse is potentially helpful to reduce the anxiety. Three children receive the injections at home for convenience/anxiety reduction.

The Specialist is normally seen every 6 months. This tends to stop after attainment of normal puberty age 13, when treatment ceases.
No reference was made to any longer term studies on the later consequences of treatment, but concerns were expressed concerning

- Impaired fertility
- Polycystic ovaries
- Early menopause
- Genetic predisposition to next generation

CHILD’S MANAGEMENT OF THE CONDITION

Nearly all the children are said to cope very well/extremely well with the condition, although many mention increased anxiety prior to the injection.

There is an ongoing concern about weight management and self image, and concern at the “loss of childhood”.

SUPPORT SERVICES FOR THE CHILDREN

A good family, close friends, and those members of the Medical Profession who demonstrated care and interest, eg “My GP has always been interested in what I had learned about the condition from the Child Growth Foundation Conference”.

A number of respondents referred to the information and knowledge they have found helpful from the CGF, the Net, the Hospital, but the majority refer to their own instincts in responding to their child’s needs.

BROADER SOCIAL ISSUES OF THE CONDITION

There is generally progressive education on the sexual issues of the condition for children of 8/9 years, with no discussion before 6/7.

GOOD PRACTICE IN CHILD’S UNDERSTANDING OF THE CONDITION

Almost all the respondents identified the benefit of honesty

- “Be straight and explain the choices of treatment”
- “Timing is important – you need to listen to your child”
- “Try not to make a big deal of it”.

A lovely quote emerged

- “We explained that she had grown faster than other children, and that after the treatment stops, it will take a few years for the other children to catch her up”.

There is some taboo associated with the condition –
“We respect her reluctance for her condition to be disclosed to her peers”.

**GENERAL COMMENTS**

“Precocious children appear older naturally and are expected to do and understand things beyond their years.

People are nasty because our daughter is a lot taller and bigger than her peers.

I struggle to explain to other children her condition – children in her class are very cruel and don’t make efforts to befriend her.

It is difficult to talk to a young child about puberty in a relevant and understandable way.

She always seems at a disadvantage, with others expecting her to be older than she really is – her childhood was somehow taken away.

There was little else we could do other than to offer our daughter tender loving care and repeated reassurance.”

**KEY LEARNING POINT**

THE SPECIALISTS/CGF COULD ASSIST PARENTS IN EXPLAINING THE CONDITION TO THEIR CHILDREN IN AN EFFECTIVE WAY, AND ASSIST IN THEIR OWN COMMUNICATIONS/EXPLANATIONS TO OTHERS

**KEY QUESTION**

HOW IS THE FREQUENCY (3, 4, 12 week) OF INJECTION DETERMINED, AND WHAT HAVE LONG TERM STUDIES DEMONSTRATED RE FERTILITY AND EARLY MENOPAUSE?
SECTION 4 - IMPACT AT SCHOOL

DISCUSSION WITH THE SCHOOL

90% of parents advise the school of the condition – the remainder are either not yet at school, or parents have chosen not to advise the school.

SCHOOLS REACTIONS TO THE ADVICE

There is a wide range of response – both positive and negative – from the School.

On the positive side:

“They have been understanding and sensitive”
“They read all the information I send them”
“School Doctor first identified the condition”
“The teachers are very good – they will inform us of any particular emotional difficulties”.

On the negative side:

“They called in the School Doctor and wanted to exclude her because of her periods and the need to change”
“She suffered abuse from a male pupil and the school’s handling of this made matters worse”
“The school was terrible – teacher said if she had wanted to teach kids going through puberty she would have got a job in a secondary school”.

HELPFUL ACTIONS BY THE SCHOOL

Schools need to be mindful to the threat of bullying and to assist the child in dealing with these situations.

Particular issues regarding swimming and changing for PE need to be managed sensitively.

It is advisable to ask a teacher to act as Mentor to assist when particular difficulties are encountered, and to act as a point of reference.

- The school should informally monitor the child’s socialisation at school and advise parents accordingly.
SCHOOL RATINGS AND GENERAL COMMENTS

Respondents marked the schools as follows:

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<thead>
<tr>
<th>Rating</th>
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<tbody>
<tr>
<td>VERY GOOD</td>
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<td>37%</td>
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<tr>
<td>GOOD</td>
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<td>17%</td>
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<tr>
<td>ADEQUATE</td>
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<td>26.5%</td>
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<tr>
<td>POOR</td>
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<td>13%</td>
</tr>
<tr>
<td>EXTREMELY POOR</td>
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<td>6.5%</td>
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GENERAL COMMENTS

On the positive side general comments included:

“The teachers allowed her (aged 7) to use their toilets to change sanitary protection”
“A teacher was designated as a point of contact”.
“The teacher called the class together and gave a talk about growth and differences”.

On the negative side

“Although the school had a letter from the Specialist, they made her life hell emotionally.”
“The treatment of our daughter (and us as parents) was appalling – I shall never forget it”.
“They thought our daughter was making mountains out of nothing”.

Once treatment starts, parents tend to minimise their discussions with the school, as pubertal characteristics stabilise:

“I try to keep the condition as low profile as possible to help maintain as normal a childhood as possible”
“I no longer feel the need to advise the school of the puberty, but they are aware she has injections every month”.

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KEY LEARNING POINT

THE CHILD NEEDS PROTECTION FROM BULLYING AND SOCIAL EMBARRASSMENT, PARTICULARLY FOR SWIMMING, GAMES, AND TOILET FACILITIES. A LOW KEY POINT OF CONTACT AT SCHOOL TO MONITOR/ASSIST THE CHILD IS HELPFUL

KEY QUESTION

WHEN THE SCHOOL/TEACHERS TAKE A HOSTILE REACTION TO THE SITUATION, WHAT CAN BE DONE TO IMPROVE THE SITUATION OTHER THAN TRANSFER?
IMPACT ON THE PARENTS

The most frequently described reactions were “devastating”, “shocked”, and subsequently “distress” and “sadness”.

The condition puts an enormous strain on the parent’s relationship, with continuing concerns about the child’s future. Over time, with treatment, increasing age, and additional knowledge about the condition, the reaction reduces, but the worries and concerns remain.

IMPACT ON SIBLINGS

Generally a mild interest from brothers/sisters. A number of parents bring brothers/sisters to the hospital when the child has his/her injection.

IMPACT ON EXTENDED FAMILY

The shock extends to the grandparents, with worry, concern, and disbelief. There is some searching for family precedents, but generally grandparents etc are supportive.

WHAT EXPLANATIONS HELP WITH FRIENDS ETC

Reactions vary from full disclosure (“I advised them what a hamartoma is and the effects it has on the child”) to more guarded responses (“I only tell close friends that the body clock started early”). One mother explains “She is a boleshy 3 year old teenager!”.

DEALING WITH DIFFICULT/VULNERABLE SITUATIONS

Parents generally encourage their children’s friendship with peers, and support their children when they are anxious. Some children become withdrawn and here parents endeavour to explain these children are not different/freakish, but rather to treat them as normally as possible.

There is anxiety regarding their sexual attraction, and the emphasis is to make clear appropriate behaviour, eg to discourage hugging and kissing except for close family, to a more direct approach – “at an early stage we had to talk about sex/paedophiles”; “try and ignore negative peer comments”.

No respondent identified particular issues, but there is continuing concern

“Am still seeking help on this myself – I think we shall see the psychologist”
“I would like some advice about that myself”
“Be confident, but don’t draw attention to yourself”.

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FIRST CONTACT WITH CHILD GROWTH FOUNDATION

This came from a wide range of sources including Magazines, through Television (Programme the Dark Secret), from hospitals including nurses in Paediatric Units, and from the Library. GP practices were not named as a source.

OTHER SOURCES OF INFORMATION

The Magic Foundation website in America, the Library, other organisations, eg the Pituitary Association.

KEY LEARNING POINT

THE CONDITION HAS A DEVASTATING EFFECT ON THE PARENTS AND IS LIKELY TO CAUSE MAJOR STRAIN BETWEEN THEM. THEY MAY BENEFIT FROM COUNSELLING OR ALTERNATIVE PROACTIVE ADVICE.

KEY QUESTION

HOW CAN THE CGF BE AN AUTOMATIC PORT OF CALL VIA THE MEDICAL PROFESSION FOR PARENTS/CHILDREN DIAGNOSED WITH THESE CONDITIONS?