

ASSERT

Angelman Syndrome Support Education & Research Trust

ISSUE THIRTY SEVEN FEBRUARY 2004

REPORT



Website: www.angelmanuk.org

Email: assert@angelmanuk.org

Telephone: 01268 415940

REGISTERED CHARITY NO. 1021882

Letter from the Chairperson

At last I have been able to get around to doing a bumper newsletter. Life as usual has been busy in the Walburn household – you all know what it's like in the run up to Christmas... Anyway, Happy New Year to you all. It's going to be a busy year for us, you will have already received details of the conference which is 3/4/5 September with this newsletter – to reserve your place please return your booking forms and deposits by 1st April at the latest – bookings are on a first come first served basis so book early!! If for some reason you have not received a booking form, please let us know as soon as possible and we will send one to you. To get a taste of a conference read the article on Page 5 about the Australian National Conference in October 2003 (no we won't mention the rugby!). Many thanks to Southern Angels for allowing us to reprint (sub group of Angelman Syndrome Australia).

ASSERT has now moved into our business premises – in expanding our own personal business, this has secured ASSERT much needed office space. It's great to have our spare room back and keeps as much ASSERT work at work rather than spending time at home on mail etc., We will still make evening calls where this is convenient to you, but are trying to keep to a 9 – 5 office timescale, we apologise if this causes any inconvenience but we hope you understand.

You should have received a questionnaire from the University of Birmingham to assist them in research they are currently doing into mood and behaviour in people with intellectual disabilities. All the questionnaires came via ASSERT, and we hope you didn't mind taking part, many

thanks for your help – we believe that research into understanding the condition is very important and may help other families in future. Hopefully you will get an opportunity to meet with Professor Chris Oliver at the conference in September.

Welcome to all the new families who have joined us – we hope you enjoy the newsletter and have a long association with ASSERT, perhaps we will meet you at the conference in September?

Finally, I hope you will take what I have to say next, seriously. We have reached a cross roads at ASSERT, we are desperate for new trustees and new blood. As you all know I have written and edited the newsletter for about six years and hope that you have all enjoyed the content of it so far. However, unfortunately I do not have the time to do this anymore – I really need to concentrate my energies on other things – including my own AS son, Matthew. Sadly this will mean that there will not be a newsletter for the foreseeable future – unless you can help! Jackie and Aleks Klimanski format and print the report, it just needs someone to type and gather pertinent information – if you would like to have a go at this please 'phone Sally Walburn. The same goes for the conference, this is the last year I will plan and organise it, so unless people step forward to support us, this event may not happen again in future. If anyone would like to discuss any issues about ASSERT or find out how you can help, then please get in touch 01268 415940.

That's all for now, please consider supporting your support group – we need your help.

Sally Walburn

Home Study Project for Carers

The Carers' Project is a tutor-supported home study scheme run by the National Extension College for carers, including the parents of disabled children. It is often impossible for parents to attend a course at the local college because of their caring responsibilities, so the project allows them to access a wide range of NEC courses including 20 GCSEs, 23 A levels, Book-keeping, Counselling, Art and Design, Writing and Basic

Skills. The flexibility of distance learning fits in well with the responsibility of being a carer as the learning can be fitted around the needs of the cared for. The project is open to carers resident in Eastern England. If you would like to know more, please contact Roz Tarry, The Carers' Project, National Extension College, The Midi Young Centre, Purbeck Road, Cambridge CB2 2HN.

Telephone: 01223 400393.

Email: rosalind.tarry@nec.ac.uk Website: www.nec.ac.uk

HELPLINE ASSERT Families/New Diagnosis/Support Line

01268 415940



Can you help?

I would really appreciate comments from families about issues, which are pertinent to Angelman syndrome since this is how we learn and can support each other. It is a real shame that virtually every time I ask for suggestions no one responds – please do this time if you can. One of our families has problems with controlling seizures in their child (not the only one I hasten to add). The Ketogenic diet has been tried, but now doctors have suggested Topirimate – has anyone any experience of this drug – positive or negative?

Also if you have had problems with seizures, what has worked and what hasn't? This parent is also keen to hear from other families where their child is severely affected by the seizures, therefore limiting their development. Remember, we are a "Support Group" so we need to support each other. Please contact the main ASSERT number on 01268 415940.

Questions and answers from the Angelman Listserve

I thought it would be a good idea to share with you some suggestions from other families which may be helpful to some of you. Unfortunately I lost a lot of other information I was going to share with you when a CD rom was wiped – don't ask me how – but I was a bit annoyed to say the least! I apologise if you don't like the term "Angel" but this is the term used in the USA.

Question:

My son has been pulling hair more at school and home lately. Actually he is sitting on the same chair as me pulling my hair as I post this! Someone mentioned talking with the school psychologist but if you knew our school psychologist – well, suffice to say, there is much more wisdom on this list. So please any suggestions?? We are trying time outs with occasional success.

Answer:

My daughter used to be the same way; we started with the phrase "hands down" and then would move her hands away from the hair and towards her own person. Then we would just get sterner with the phrase, and eventually 'smack' the top of the hand doing it. while saying NO ..Hand Down! She rarely pulls hair now...I did not let them smack her hand at school but they did use the Hands Down phrase with authority. The most important thing with an Angel is to remember NOT to smile back when you are trying to teach them something. Remind the school of this too. The angel will use their smile for manipulative purposes my daughter will reach for hugs now, but not by pulling hair, now she just puts you in a headlock! But hey, it doesn't hurt like pulling hair!

Question:

Hair pulling, pinching and biting have become a "huge" issue with my daughter. I'm at a loss as to what to do about it, and it is affecting everything she does and everyone she is around. I've tried as much as I can think of, all to very little avail. Ideas?

Answer:

We've also had loads of trouble with our daughter and pinching, biting and hair pulling. She used to grab people's hands and bite them, but after a LOT of work and redirecting, she still grabs people's hands but now she kisses them. She still pinches (the bigger she gets the harder the pinches too, I have the bruises to tell) and hits and pulls hair when she is fed up, (I got a lovely note from her EPA today that she was quite aggressive at school today). Our only way of dealing with this right now that works most of the time is trying to see in her mood that maybe something is coming and to back off, out of reach, and make sure she is sitting in class where she can't reach anyone (because sometimes there is no rhyme or reason for the swing/pinch /whatever). She doesn't like to be excluded from activities, so removal from situations sometimes works too; she knows she has to be "good" in order to participate. On the other hand, she also is smart enough to use bad behaviour as a way of getting out of things she has to do, so we have to see it for what it is sometimes, and keep plugging on whether she likes it or not. I do remember reading somewhere that these are typical behaviours for Angels, also "age appropriate" as she is dealing in the 2-3 year old level with frustrations, etc.

Suggestion:

I don't know if this helps but when our daughter gets suddenly aggressive (hair pulling and hitting anyone near her), we have found it to be either a sugar rush (from something she has eaten including food colourings which can set her off – red and blue) or she needs food or she is overwhelmed by lots going on around her (too much activity to take in). We have found her calmest days are when she is fed snacks a lot during the day and lower social activity. For a while there I was thinking she might have diabetes but a quick blood test at the Dr's disproved that. We just observed the behaviour and took notes at the time, what was happening, what had she eaten or drunk or when did she eat last etc.

Contents of the Newsletter

You will notice that the pieces in this newsletter are drawn from a wide source of places. This is tremendous, and we hope you will agree that it makes interesting reading. However, while we are keen to promote discussion and to pass on many views and experiences, it is also important to appreciate that the opinions and views expressed by contributors to this newsletter are personal ones and not necessarily those of the trustees of ASSERT.

Please note that the deadline for the next edition of the ASSERT Report is 30th April 2004.



Things to ponder

Can you cry underwater?

How important does a person have to be before they are considered assassinated instead of just murdered?

If money doesn't grow on trees then why do banks have branches?

Since bread is square, then why is sandwich meat round?

Why do you have to "put your two cents in"...but it's only a "penny for your thoughts"? Where's that extra penny going?

Once you're in heaven, do you get stuck wearing the clothes you were buried in for eternity?

Why does a round pizza come in a square box?

What did cured ham actually have?

How is it that we put man on the moon before we figured out it would be a good idea to put wheels on luggage?

Why is it that people say they "slept like a baby" when babies wake up like every two hours?

If a deaf person has to go to court, is it still called a hearing?

If you drink Pepsi at work in the Coke factory, will they fire you?

Why are you IN a movie, but you are ON TV?

Why do people pay to go up tall buildings and then put money in binoculars to look at things on the ground?

How come we choose from just two people for President and fifty for Miss America?

Why do doctors leave the room while you change? They're going to see you naked anyway.

If a 999 operator has a heart attack, whom does he/she call?

I signed up for an exercise class and was told to wear loose-fitting clothing. If I HAD any loose-fitting clothing, I wouldn't have signed up in the first place!

When I was young we used to go "skinny dipping," now I just "chunky dunk."

Wouldn't it be nice if whenever we messed up our life we could simply press 'Ctrl Alt Delete' and start all over?

Stress is when you wake up screaming and then you realize you haven't fallen asleep yet.

My husband says I never listen to him. At least I think that's what he said.

Why is it that our children can't read a Bible in school, but they can in prison?

If raising children was going to be easy, it never would have started with something called labour!

Brain cells come and brain cells go, but fat cells live forever.

Child Tax Credit

If you are receiving Child Tax Credit with a gross income of less than £13,230, children can claim free school meals.

Car Safety

Safetex Ltd supplies a range of harnesses for children and teenagers that are fitted outside the vehicle and then clipped to the removable fixing straps on the seat. Tel 01372 451 272 or visit www.safetex.com

Newspaper contact in Harlow

If anyone is doing any fundraising activities in Harlow, Bishops Stortford, Epping areas, please contact Tracy Hubbard on 01279 624352 or email: tracy.hubbard@archant.co.uk – where you will receive free publicity.

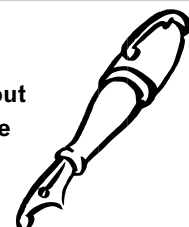
Holiday Accommodation Guide

Radars' guide for disabled people to holiday accommodation in Britain and Ireland is now available on line. You can pick the area you are interested in and find out information related to your requirements, including special diet, bedding, hoists, parking, no smoking etc., www.radarsearch.org/welcome.asp

New Holiday Centre

Accepts families including those who want periods of respite support for children during the holiday. Clynyfw, Abercych, Boncath, Pembrokeshire SA37 0HF
Tel: Jim Bowen 01239 841236 – direct payments can be used to fund visits. For Carmathenshire residents there is a voucher scheme allowing people with a Carers Assessment to use local authority respite funding, and it is hoped that the scheme will eventually be extended to residents of other areas.

If you enjoy these articles please think about writing one about your family, we really find this the most interesting part of the report. The address to write to on this or any other subject is: ASSERT, PO Box 13694, Musselburgh, EH21 6XZ



Break

www.break-charity.org Affordable self-catering holidays for families with special needs in Norfolk.
Tel: 01263 822161
Fax: 01263 822181

I Saw A Child ...

I saw a child who couldn't walk,
sit on a horse, laugh and talk.
Then ride it through a field of daisies
and yet he could not walk unaided.

I saw a child, no legs below,
sit on a horse, and make it go
through woods of green
and places he had never been
to sit and stare,
except from a chair.

I saw a child who could only crawl
mount a horse and sit up tall
Put it through degrees of paces
and laugh at the wonder in our faces

I saw a child born into strife,
Take up and hold the reins of life
and that same child was heard to say,
Thank God for showing me the way.

*"The real voyage of discovery consists not
in seeking new lands but in seeing with
new eyes" Proust (1871).*

New National Help Line

Purple Pages – telephone line to help carers and people with disabilities to locate services.
Run by the Care Forum 9.30 – 4.30
Monday – Friday
Freephone 0800 808 5252

Talking Point

A website to help doctors, teachers and parents offer support to children with speech, language and communication difficulties has been launched –
www.talkingpoint.org.uk

Second Space

Second Space is a new charity aimed at providing parents of children with special needs with a weeks break. These breaks will take place in beautiful second homes donated by the owners, to the charity, for one week in every year. The home will be offered for free and the charity aims to cover the cost of transport through sponsorship.

For families where alternative respite care for the special child is available for the period of the holiday, the idea is to offer the parents a complete break from their caring role and where there are other children, to enjoy some stress free time with them. If respite care cannot be arranged, the charity will consider offering the whole family a holiday where possible, although the homes used are not equipped with any special facilities that some families may require.

Second Space will have in the region of 50 homes on their books, both in the UK and abroad for 2004. If you would be interested in one of their holidays, you can call them to request an application form on 020 7792 9043 or write to them at Second Space, 6 Chepstow Villas, London W11 2RB.

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A Parent's Guide to Direct Payments

This is a new publication from the Department of Health to coincide with a change of the regulations in England from April 2003. In the past parents could not insist on direct payments, which were made available at the discretion of the local authority. If allowed, they could be used for any community care service, which the local authority has assessed you as needing and provided for care to be "bought in" rather than provided directly by local authority services.

However, now, in England, a request can only be refused in very limited circumstances, but must still relate to a service, which Social Services have assessed as being needed.

The booklet goes through the whole process from explaining what a direct payment is and how it can be used, to the procedures for organising a service. Copies are available from Department of Health, PO Box 777, London SE1 6XH. Fax: 01623 724524. Tel: 08701 555455.

Local authorities in Scotland must also now offer direct payments to all who are eligible.

Guide for Disabled People or Carers

The Department of Health has published a new edition of its practical guide for disabled people. The guide is aimed at disabled people, older people, people with a mental health problem and those who support or care for someone. It provides details of how to find or obtain information, services and equipment – it is available free from Department of Health Publications, PO Box 777, London SE1 6XH Tel: 0800 555777 Fax: 01623 724254 and www.doh.gov.uk/disabledguide

ASA 6th National Conference Sydney 3 – 5 October 2003

We arrived in Sydney at lunchtime after a smooth flight. We took a taxi to our motel where we relaxed until it was time to go to the pre conference drinks at the St George Leagues Club. It was wonderful to catch up with old faces, meet new families. After drinks we had a lovely tea with our old friends Julie and Barry Hunt (their daughter Sarah was the first AS child diagnosed in Southern Australia) and a couple from New Zealand.

SATURDAY

Simone Kennedy (President) welcomed everyone. Simone spoke of the past 10 years of the Association and its achievements:

- AS Brochure
- T-shirts, Badges
- 60 minutes interview
- Ray Martin Show
- Establishment of AS Clinic in Sydney
- National Newsletter
- Five Conferences
- Information package
- Social Events
- Network of support between families Education of wider community
- Web site
- Prominence presence on board of International Association

States represented were NSW, SA, QLD, VIC, ACT, Tasmania and a family from New Zealand. Approximately 70 attended the conference day.

It was lovely that the Conference was dedicated to Jenny Sims (Parent and ASA Board member) and Alyssa Hannaford, AS daughter of John and Robyn Hannaford. Alyssa passed away on 16 September 2003.

Kevin Kennedy presented a wonderful photo display of all Conferences over the past ten years.

Dr Ellie Smith – International Proceedings:

Ellie spoke on the 2nd IASO World Conference and the 8th ASF Biennial Conference, which was held in Washington, July 2003. 360 attended the majority being from the USA. 52 from outside countries, 1 from Australia. 19 countries represented in all. Every aspect of Angelman Syndrome was covered. Of interest to Ellie was the following:

- Assistive technology in the home. Ellie spoke about a computer system that was geared for the child with AS.
- The Angelman Syndrome Project, Louise Tiranoff from USA spoke about the web site she is developing – is currently in-putting all data on AS on a database. The aim being that interested people can access information about AS via the internet, which will assist parents and professionals.

Genetics:

Ellie spoke briefly on the genetics of AS. Ellie covered all the different AS mechanisms involved. Also the different tests that can be done to determine the type of AS.

Ellie spoke about a particular case of a woman with Prader Willi Syndrome adult having an Angelman Syndrome baby. Ellie

explained that this case provides absolute proof on how the genetics of AS and PWS are inter-related.

Dr Charles Williams (USA) and Jill Clayton-Smith (UK) spoke on the diagnosis process of AS (reports are that all is still the same).

The gene for Retts Syndrome has now been found. People with suspected Retts could now be tested for the diagnosis.

Betaine/ folic Trial – Dr Art Beaudet (USA) talked about the increase of methylation in the brain of AS children. Dr Beaudet completed this study in an attempt to determine whether there was any improvement in IQ, behaviour etc. He recruited 20 AS patients, measured IQ, behaviour, communication, walking, sleeping. The results were that nothing had changed (some children were given a placebo). There were lots of questions on how folic acid in bulk three times a day works on the male chromosome. The study was generally found to be inconclusive, but they are continuing work in this area.

Neurology and Epilepsy management – Dr Jayne Anthony/ Dr Don Pryer (Consultant to St George Hospital, Sydney)

Dr Don Pryer spoke about the different types of seizures people with AS have, also the different types of drugs used in epilepsy

Phenobarb: Favourite – sedative, cheap and reliable, oral or injected

Dilatin: Still used a lot, some don't due to side affects (is hard to get dose right, too many side affects).

Tegretol: Quite common, tablet form, is suggested that sometimes worsens fits in AS.

Epilim: Most frequently used drug top of the pops, has side affect of weight gain however usually not seen in AS.

Lacnictal: Most popular 10 years ago.

Rivotril: Sedative, but used a lot.

Zarontin: Used for particular types of epilepsy

Valium: Sedative good for emergency situation.

Frisium: Variant of valium.

Midazolam: Sedative drug.

Galoitril: New drug.

Trileptal: New drug.

Keppra: New drug.

Dr Jayne Anthony – Spoke on management of epilepsy in the home. Early Treatment may decrease mortality and morbidity. A delay in treatment testing lessens the likelihood that a single medication will abort seizures. Talked on methods of acute treatment of epilepsy by carers:

- Rectal Anticonvulsants.
- Buccat Midazolam – Which are ampoules that you squeeze contents into side of cheek. Studies to date show that there is not much difference from time of administration to the end of the seizure. Advantages are that it is easier to administer, is more acceptable in public and preferred by carers who administer medication. Suggested that a dose be given a GP's Surgery or Hospital setting to monitor any side effects. Generally if a seizure exceeds two minutes preparation for administration of buccat midazolam or rectal diazepam should be started. Of course each child varies and seizure history is invaluable in deciding plan of action for GP. Buccat

midazolam is bought in hospital pharmacy only. A nurse from hospital will instruct the Carer how to use it. Cost is minimal as you can use a Health Care Card at hospital.

Ask The Panel:

The Panel consisted of:

Dr Jim Papadopoulos

Dr Robert Leitner

Dr Ellie Smith

Keith Bradley – Parent

This was an opportunity for parents to ask individual questions about their child to the panel.

Keith Bradley as always was there to offer his words of wisdom as only Keith can. A parent who was having difficulty with her AS child's sore eyes asked a question. Keith offered that his son Callum has a problem with fluttering his eyes, however, Keith stated this was "a form of seduction" on Callum's behalf. This comment brought the house down!

Sleeping with Angels Dr Jim Papadopoulos

Dr Jim Papadopoulos is a specialist in Pediatric Sleep Medicine. Spoke about sleep apnoea, symptoms and behaviours that arise and how this can be harmful.

Completing a sleep study on children with a disability can be difficult. A sleep study measures sleep, how it can be treated and determines high risk groups. Children with OSA have similar behavioural problems to children with ADHD – hyperactivity and aggression was found to be increased in those who snored. Generally, not enough melatonin in the brain causes lack of sleep. The melatonin levels drop during the onset of puberty. Teenagers will generally want to go to bed later and later. Only one study on melatonin has been done on children with AS. Thirteen children were involved in the study. The findings were that 11 were helped if they increased their melatonin levels Dr Papadopoulos believes that we need more research into sleeplessness in AS. The impact is not only on the child but also the entire family. Spoke of Behavioural Sleep Disorder. He explained that when your child goes to sleep with their dummy for example, when they partially awake, if there surrounding environment has changed (i.e. dummy missing) then they wake up. He suggested you replace dummy with a soft toy, soft blanket or night light or larva lamp (if that is possible with our AS kids!). Spoke on sleep management – need strict bedtime rules, organic problems need to be managed i.e. illnesses, asthma etc. Sleep Hygiene – do not hype up before sleep, quiet, dark, try controlled comforting if child awakens, having a sleep diary helps. Dr Papadopoulos indicated that he was keen to do further research on melatonin and AS. Dr Papadopoulos has welcomed any questions be forwarded to: Dr.djp@optusnet.com.au

Interesting Information: Generally the medication Epilim reduced melatonin secretion which may slow down the onset of puberty.

A parent had a good idea in how to get their child to fall asleep at night. They found that by giving their AS child two dummies at night, the child put one in his mouth then spotted the other and put that in his mouth, the constant movement helped their child fall asleep.

Adult Angels

Mrs Mary Bills – Parent.

Talked about her son Michael who is 19 and has finished school. Mary spoke about the reduced services once Michael has finished school and what services are offered for her son – respite, leisure activities etc.

Siblings Needs

Mr Jurgen Willie – Social Worker

Spoke on the grief and loss process and how this relates to having a child with a disability. Spoke on disenfranchised grief, which is grief that cannot be socially shared. The effect of this is the lack of social support, which relates to siblings. Parents then spoke openly about their own experiences with their siblings acceptance/non acceptance of their AS sibling. Jurgen suggested having open communication between parents and siblings, allow them opportunities to express their feelings.

Communication

Ms Ylana Bloom – Speech Pathologist

Ylana gave a very expressive and passionate speech on communication. She spoke on technology and communication:

<i>Low Tech</i>	<i>High Tech</i>
Compic	Computer generated
Unaided/ aided	Digitised
Sign language	Big Mac

Need to consider the value of some high tech devices for use in the home, however, in a school setting more appropriate. Where receptive language is greater than expressive language need to have augmentative communication to bridge the gap. Ylana suggested that the auditory memory problems our children have may be due to medication and seizure activity which has a huge impact on day to day performance.

Ylana spoke briefly on about PECS – advantages/ disadvantages. Advised that it is necessary to assess each individual child to determine appropriate communication system for them. Ylana has developed her own communication system by using picture symbols

Thank You: Simone Kennedy (President) gave thanks to the organising committee. Heather Church (VIC State Vice President) gave a heartfelt thank you to Simone Kennedy for her involvement as President of the National Association for the past nine years.

Saturday, Conference Dinner:

Dinner was held at the St George Leagues – Club in Kogarah. We had a lovely dinner and was entertained by a 20 minute video of the – last 10 years of Angelman Syndrome. My highlight was seeing Grant's mullet again – (they showed the "Today/Tonight" segment that aired in 1994). It was laughs all around with how we all looked only 10 short years ago.

Keith Bradley again had his mandatory After Dinner Speech, which was full of interesting and amusing stories of his son Callum's continuing quest of acceptance when socializing in public with of course the great assistance – and support of his father Kevin! There was also a very successful auction with Kevin the Auctioneer and Barry Hunt his lovely assistant.

Sunday – AGM/Family Picnic Day:

Annual General Meeting:

The AGM was well attended. Simone Kennedy chaired the AGM, this being her last official duty as President.

This day is my favourite part of the Conference weekend. This is the day I get to meet – other AS children and adults. We had a wonderful time for the short period we were able to stay (we had an afternoon flight out of Sydney). The picnic day was to be attended by approx. 140 people. The weather was fine with a little cloud cover. I was told later that a downpour arrived about an hour after we left. So that was our journey to Sydney, I hope you enjoyed this report and that you may have gained a little more knowledge from this Conference. I would like to

thank Simone, Kevin, her family and the organising committee for all the little extra's they helped us with. Also to Qantas for getting us home safely!

Thank you to IDSC for the financial support which greatly assists families being able to attend these interstate conferences.

Many thanks to Kerri Monaghan for her report – it's good to catch up with our down under friends and families – we don't know them personally but the common bond of Angelman Syndrome unites us all!

The Green Paper: Every Child Matters

Published in September, the Green Paper is a consultation document that sets out government proposals for the direction of children's services, following the inquiry into the death of Victoria Climbié. The aim of the Paper is to promote integration of services and clear lines of responsibility and accountability so that children, whether at risk or otherwise, do not fall between any gaps in services.

The Paper identifies ways to promote children's rights and to protect them in settings including Health, social services and education, so that they are able to maximise their potential. The Paper notes that many children are seen by a number of different professionals, but with no central register or integrated working between agencies, any concerns about their welfare were not being flagged up and shared.

How the Paper intends to improve services:

- Supporting parents and careers;
- Early intervention and effective protection;
- Accountability and integration – locally, regionally and nationally;
- Workforce reform.

The key proposals of the Green Paper are:

- Children's directors in every local authority to be responsible for children's social services and education;
- The appointment of a children's commissioner for England;
- Children's services brought together under children's trusts;
- Multi-agency teams based in schools and children's centres;
- Local safeguarding children boards to replace area child protection committees;
- Children's services to be inspected on joint working through an integrated inspection framework overseen by Ofsted;
- Development of a unique number for every child to help electronic information-sharing among agencies;
- Establishment of a children's workforce unit.

Many of the proposals will be of direct benefit to children with intellectual and physical impairments and their families, particularly those aimed at improving information sharing between agencies, developing a common assessment framework, encouraging more families to use the direct payment scheme and introducing a lead professional to ensure a coherent package of services to meet the individual child's needs.

In the long term it is planned that children's services should be brought together under the Director of Children's Services to form Children's Trusts. There will also be an obligation on local authorities to work closely with public, private and voluntary organisations to improve outcomes for children. This local partnership will be supported at government level by the newly created post of Minister for Children, Young People and Families in the Department for Education and Skills (DfES).

There will also be a duty on local organisations (such as health) to work together with other organisations to safeguard children and promote their wellbeing. The document sets out proposals for a new Children's Commissioner to act as an independent champion for children.

Although much of the above should be heralded as a commitment to improving integrated working practices; there are some concerns about the Paper's proposals (or lack of them) for children with health needs. It is proposed that education play a prime role in the co-ordination and inspection of services and there are fears that the needs of children with disabilities and long-term or life-limiting health conditions would not be accommodated very well in these circumstances. The Green Paper devotes relatively few paragraphs to the complex needs of these children and it must be made clear during the consultation process that children with medical needs must not be overlooked.

There is also some cause for concern regarding the requirement that service providers appoint staff with a specific child protection remit. This will have a big resource implication for many children's services.

To see the Green Paper go to www.dfes.gov.uk/everychildmatters

We are drowning in information but starved for knowledge – John Naisbitt

Coping with the system

The following article is taken from a series of articles covering topics relevant to carers, by the Princess Royal Trust for Carers: Many carers find that the 'system' isn't an organised system at all and the range of services available varies hugely from area to area. One way to help piece together a jigsaw of services is to keep a diary or notebook and write down the date and details of any contact, that you have with the professionals.

Useful questions to ask: Who are you? Which part of the service do you work for? Where are you based? Can I contact you directly if I need to? How do I contact you? Is there a best time of day to get hold of you? What do I do if I need help outside office hours?

People sometimes find it difficult to get the help they need from professionals. Contact can sometimes be frustrating and even distressing. Build up your confidence, it helps if you can feel confident in yourself and are clear about what you need. Remember that confidence increases with experience. Draw on your own experience. Think back over some of the difficult things you've done successfully – encourage yourself by recalling these positive experiences.

Develop a positive attitude. If you feel confident and believe in yourself and in the value of what you're doing, the people you come into contact with will value and respect you in turn. Be clear about what you want. Be specific – think about the important facts that the person you're speaking to needs to know. Say what you want directly, you don't need to be apologetic about saying what you need. Stick to what you want to say and don't be side tracked.

Sleepsuits

The following information came from Lynn Scowcroft who uses the sleepsuits for her son Jamie.

The All in One suits come from a catalogue (not special needs) Vertbaudet tel: 08452 700270. I use the fleece all in one's with Jamie in winter – due to him kicking his bedding off, I hope this will be useful to other readers. I think they are a reasonable price and wash very well, and if need be can be worn back to front. Prices are from £8.99 for a 2 year old to £10.49 for an 11/12 year old (the largest size) Cotton and for the fleece is £8.99 (age 2) to £11.49 (age 11/12), they come with or without feet.

Conference 2004

- When?** Weekend of 3-5 September 2004
Where? Loughborough University, Loughborough, Leicestershire
For whom? Families/ Carers of children/adults with Angelman Syndrome
How Much? £75 per person for the weekend
£40 for a child under 12 years for the weekend
FREE to all with Angelman Syndrome
FREE to your carer for the person with Angelman Syndrome

Programme to include: Respite, Transition, Behaviour, Seizures, Genetics, Communication, Education, product displays and anything else we think will be of interest to you and we can arrange!

All this is included in the price:

Accommodation
Childrens Outings/Activities
All meals/special diets catered for
Entertainment
All of the conference talks and coffee breaks/conference lunch
Conference packs

Can you afford to miss it?

Once again we have had a few individuals who have generously donated sponsorship and fundraising money to the conference which allows us to reduce the cost substantially. Places will be allocated on a first come first served basis so when you get your initial forms please reserve you place.

As last time if a parent comes as a parent/carer i.e. you

look after your own son/daughter/siblings for the duration of the conference – you can come free – this does allow you or your partner to swap, for example Mum goes to the conference on Saturday, and Dad cares, then on Sunday Dad attends the conference and Mum does the caring, you will still have one free place. If however, you bring an additional carer (over the age of 18 years, please) which allows both of you to attend the conference you will have to pay for both of you but not the additional carer – we hope this explains the position but if you do have any queries on this please check with us and we will be happy to clarify this for you.

The conference will be extended to Sunday afternoon this time or we can start earlier on the Friday – if anyone has any contribution to make or suggestion or can help in any way please contact ASSERT. If you would like to be part of the organising committee we would be really grateful for your support.

DON'T MISS OUT !

The conference is a great way to network and make friends and learn from other AS families as well as the experts – we would say that everyone who attends the conference doesn't want to go home at the end – and wants to come back again. It is very relaxed and there is no pressure – we can all be ourselves and we don't have to make excuses or worry about how our children/adults are going to behave – we are altogether and know what life is like with Angelman Syndrome. It will be great to meet with you and see you there – we are looking forward to seeing as many of you that can attend! Please reserve your place.

SEE YOU IN LOUGHBOROUGH!

Websites

www.pediatrics.about.com/cs/specialneedskids/

Sensible day-to-day information on care questions and techniques for parents and carers of children with special needs, collected together by a doctor.

www.comeunity.com/disability/

contains useful information, book reviews and links for special needs, put together by an American parent support organisation. Includes [/seizures.html](#); [/speech/index.html](#) on speech and communication; [/cerebral_palsy/hemiplegia.html](#) on milestones and activities for toddlers with cerebral palsy; and [/sleep-links.html](#) on sleep difficulties in children.

A related section

www.comeunity.com/premature/child/growth/index.html is on growth and feeding issues of children.

www.mothenature.com/Library/bookshelf/Books/50/111.cfm

Very full resource for safety, first aid and hints for caring for children at home. The author has thought of almost anything that could happen, under 148 headings. The first aid subjects include clear guidelines for when to take a child to a doctor. http://topics_a-z.parenthood Pregnancy and "parenting" topics including some more unusual ones to see covered, such as transitions, family relationships, travel and ideas for celebrations.

www.spechtx.com/cpalsy/cp.htm

Joint effort by an occupational therapist and a speech therapist on early intervention techniques for cerebral palsy. Includes outlines on adaptive equipment and positioning, sensory experiences, augmentative communication, feeding issues, and adaptive computer technology.

www.pediatrics.wisc.edu/childrenshosp/sicknewborn/t-devp t.html

For parents going home from hospital with a sick baby, straightforward suggestions for age-specific activities to encourage development up to three years old. Compiled by a hospital centre for perinatal care.

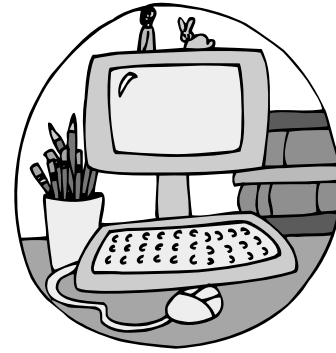
Scoliosis – Two sites that taken together, are informative about scoliosis, a curvature of the spine, and the surgical techniques that are sometimes used for it. www.scoliosisrx.com and www.jdryerscoliosis.com/View_Procedures/view_procedures.html.

www.techconnections.org/forum/index.cfm

A consumer site for assistive technology. Site independent of the suppliers of these products, where users of the products can report back for others, the pros and cons of particular items of equipment.

www.24dr.com/index.asp

Brief medical encyclopaedia and dictionary of medical terms, with some other general health resources.



ASSERT Website

www.angelmanuk.org

ASSERT Email

assert@angelmanuk.org

www.radarsearch.org

Complete, free on-line version of the RADAR (Royal Association of Disability and Rehabilitation) annual guide to accessible places to stay. Detailed descriptions of what facilities are provided by each one, and general holiday-type information about each region of the UK.

www.accessart.org.uk

Collection of free, visually innovative on-line workshops in educational arts activities for children and teachers.

www.drugs.com

American drugs site with an interactions checker. Includes interactions with other drugs and with foods and drinks. Note that as the site itself says very clearly, there are some differences from drugs in the UK, but it may still alert to something that has been missed.

www.europeanchildrensnetwork.org

Site of a coalition of European organisations that are working to promote the interests and rights of children in EU policy. Children can join the on-line forum to discuss their views on how policy should develop in Europe to benefit them. The organisations can then take up their views to try to influence policy as it forms.

www.tiger.gov.uk

Department of Trade and Industry site with a user-friendly guide on the minimum wage, maternity and paternity rights, adoption and flexible working rights in employment. (Tailored Interactive Guidance on Employment Rights)

www.probonouk.net/Resources

for those potentially giving or receiving free legal help. It does not carry names of practitioners giving free help but points the way to obtaining it, under legal aid or pro bono schemes.

Speech Therapy and the Statement

The following article, was written by Sally Capper of the Down's Syndrome Association, first appeared in Network 81's newsletter.

Who is responsible for speech and language therapy?

It is part of the remit of the health authority to provide speech and language therapy services and indeed most speech therapy is provided by the health service.

The local education authority (LEA) will usually ask the health authority to provide speech therapy in an individual case but the health authority can say no. It is lawful for them, the health authority, to rely on a shortage of funds to say it would not be reasonable to comply with the request.

This contrasts with the position of the LEA. If the amount of speech therapy your child needs is clearly set out in Part 3 of his or her statement of special educational need as special educational provision the LEA cannot escape its legal responsibility to arrange that it is put in place. The LEA can try to arrange the speech therapy by asking the health authority but if they do not agree to provide it then the LEA must arrange it some other way. They have a personal legal duty owed to the child under the Education Act 1996 S.324(S)(a)(i).

Because the LEA cannot escape the duty once the speech therapy is specified in Part 3 of the statement, it is common for LEAs to take steps to make it difficult to get the speech therapy clearly specified. They also often suggest that it is non-educational and should appear in part 6 of the statement.

The Speech Therapy advice for the Statement

Because the contents of the statement of special educational need will be based on the professional evidence, the advice from the speech and language therapist prepared for the statement is very important. It should include a clear specification of all the therapy necessary to meet your child's special educational needs and not just a description of the needs.

Sometimes speech and language therapists are under the impression that they are not allowed to include the amount of therapy needed in their advice. This is not so. Paragraph 7.79 of the Code of Practice states that "LEAs should not have blanket policies that prevent those giving advice from commenting on the amount of provision they consider a child requires."

The advice may suggest that your child's needs can be met by a language programme carried out in the school. Even if this is the case you will still want it to specify how frequently the therapist should attend the school to see how your child progressed and update the programme.

Getting the therapy in Part 3

The leading case, deciding that speech therapy can be special educational provision, commonly referred to as the Lancashire judgement, took place in 1989. Despite this, LEAs are still arguing that speech therapy cannot be special educational provision because it is usually provided by the health authority. Not only can speech therapy be special educational provision and be properly placed in Part 3 of the

statement, but it is only in the rarest cases that it will not be so. The Code of Practice at Paragraph 8.49 makes this clear by saying "since communication is so fundamental in learning and progression, addressing speech and language impairment should normally be recorded as educational provision unless there are exceptional reasons for not doing so."

If the speech therapy is set out in Part 3 your child's right to it can be enforced, if necessary by legal action. If, on the other hand it appears as non-educational provision in Part 6 of the statement no-one has any responsibility to provide it.

Wording to avoid

Some LEAs habitually use wording in the statement that is vague, unspecific and likely to lead to confusion. The following are to be avoided.

Ambiguity over who is to do what. For example "Two sessions of speech therapy weekly". A parent might expect a therapist to attend while the LEA was intending to rely on the LSA. Make sure the statement requires that speech therapy is to be carried out by trained speech therapists if this is what you believe is necessary for your child.

Ambiguity over frequency of provision. For example "Regular speech therapy" which could be once a year or even less.

Ambiguity over the amount of provision. For example "up to four hours speech therapy a term". This doesn't guarantee any level of therapy at all for the child. Ask for the exact amount or at least the minimum to be specified.

Provision that is limited in time despite the fact that the child's needs will continue. For example "10 sessions of weekly speech therapy". It is always open to the LEA to amend the statement if they are advised that a change in provision is required but this wording leaves the child with no provision after 10 weeks.

Wording that allows someone else to decide about provision. For example a statement should not say, "speech therapy as advised by the speech therapist". Of course the current advice of professionals should be considered but it is for the LEA to make a decision about provision in the light of that advice.

What the Statement should say.

You will need professional advice to decide your child's speech and language therapy needs. If you are unhappy with advice from the health service therapist or they cannot persuade you to specify what is needed you may want to consider paying for a private speech therapy assessment of your child.

The following is just an example of what the statement might say.

"Deborah needs a language programme to be carried out by her classroom assistant at least three times a week but preferably daily in two short (20 minute) sessions each day. The programme should be drawn up, supervised and monitored by a speech therapist with knowledge and experience of children with Down syndrome. The speech therapist should attend the school for a one hour session every fortnight in the autumn term

to demonstrate to the classroom assistant the strategies for delivering the programme and to assess Deborah's progress and update the programme. The following term and thereafter the speech therapist should attend the school for a one hour session at least twice a term for the purposes set out above."

**National Audit Commission
Children with Disabilities and
their families**

If you need to substantiate any arguments for provision, you may like to use the information gathered by the National Audit commission in their report in September 2003 – it demonstrates problems parents and families encounter over the years dealing with services and the lack of provision, for more details have a look at the website on www.audit-commission.gov.uk/disabledchildren

Mr. Jacob the Angelman

This poem was written by Michael a cousin of Gloria Sholtz (USA) – thanks for her permission to print.

My cousin Jake is a boy that's five years old,
Whose lessons to me are worth more than gold.
He struggles with simple tasks every day,
But he always seems to find his way.
Our family was deeply heartbroken and torn,
For with a disability he was born.
The thing that Jacob is all about,
Is never taking the easy way out.
His persistence is as hard as nails,
He will keep on trying until he prevails.
I will take things for granted no longer,
For this one person's life has made me so much stronger.
With each small step that Jacob takes,
And with every accomplishment that he makes,
I will be there by his side,
Like a lighthouse, to be his guide.
Jacob will never speak for sure,
But his smile will open every door.
I cannot imagine how my life would be,
If Jacob were not here to help me see,
How someone so small and challenged could teach,
That there is never a star that is out of your reach.

Direct Payments

The extension of direct payments to 16 and 17 year olds has been investigated by the Joseph Rowntree Foundation. Young disabled people thought that direct payments could be a good way of achieving more independence, but could sometimes change relationships with parents. An independent living scheme, for example, can support young people managing direct payments.

To find out more: www.jrf.org.uk/knowledge/findings/socialcare/553.asp or order the report from York Publishing Services on 01904 430033.

Fundraising

Many thanks to everyone who has been fundraising for us in many ways, there has been money raised by Scouts, coffee mornings, sales, Christmas card sales (Web Ivory), marathons (Windsor Half Marathon), wedding anniversaries – congratulations to Margaret and James, grandparents to Kieran Buchan, they celebrated their 30th Wedding Anniversary, donating money which they received in lieu of presents...collection boxes, stamps, paper, and many other ways – this is brilliant and we are very grateful, some has been specified for the conference so the coffers for that are growing which is great!

Bernie Silver represented ASSERT to receive a cheque for £300 from the 57th Anniversary service of the 18th Edmonton Scout Group on 2nd November 2003. Bernie wrote:

It was with great pleasure that ASSERT was invited to attend the service by Colin and Sue Bateman. While at the service we were presented with a cheque for £300 from all the Beavers, Cubs and Scouts, from all their efforts fundraising over the past year. This is the second year that they have fundraised for ASSERT. Paul Bateman who has AS is a member of the Cubs but will be moving to the Scouts shortly. It was great to see how they have taken Paul into their group and just treat him as one of the Pack. Thanks to all who have raised funds and donated so generously to ASSERT.

Here is the hall of fame
ASSERT Donations received up to
and incl 07.01.04

E A Abrams
Mrs L K Anderson
Asda Stores Marsto, Blackpool –
Colleague Circle
Barkingside Rotary Club
Nicki Barrasse
Bishop Douglas School
L Buchan
C & GC Charitable Trust
Dr C Cohen
Joan Copen
Crossed Keys Masonic Lodge,
Canvey Island

Gareth Davies
18th Edmonton Scout Group
Mr K C Goodwin
Lassinder
Mrs J McCulloch
Patriots Scooter Club
Karen Robertson
Mrs Scowcroft
Mr and Mrs Simpson
Matthew Swallow
Mrs H M Tait
J Thornton
Ken Walburn

Regional meeting at Mawdsley (North West Region)

A Regional meeting at Mawdsley (North West Region) has been organised by Rachel Edgar and Rachel Simon on Saturday March 6th 2004 10.30 – 4.30 p.m. If you have received information already, please return your forms as soon as possible so that we can ascertain numbers. If you are not in the region but would like to attend, please contact Sally Walburn on 01268 415940, to see if there are any spare places available.

Hurtling into a void?

If you are a parent of an older child you may be interested in reading this article that was written by Madeleine Cowley using her own experiences when transferring her son to adult education. Madeleine is a trustee of Network 81 and this is part of an article that first appeared in the Network 81 newsletter.

Thanks to the SNAP Brentwood newsletter for bringing this to our attention.

The teenage years are often very difficult ones for young people and their families. Decisions have to be taken and moves made that can have long-term consequences. For young people with special needs this is a particularly stressful time. The opportunities and choices may not be as evident and accessible as they are for their siblings. For their parents this should be a time when they begin to 'let go' and see their child move – into adulthood, but for many the reality is that the pressures grow and their caring responsibilities increase.

This time is now recognised as 'transition'. The SEN Code of Practice sets out what should happen in schools. The Connexions Service has a remit to provide advice and guidance to young people with disabilities up to the age of 25. The recent White Paper 'Valuing People' sets out responsibilities for transition issues and emphasises the need for effective links between children's and adult services. I could go on... BUT what is it like for young people and their families in reality?

Although some families have good experiences, the majority of parents feel completely unsupported and are left to search for information about options and possible provision themselves. The transition process is complicated, involving several different agencies. Parents and students need to understand the process – what happens, when, who is involved – and be able to identify a 'key worker'. The current system is confusing, jargon laden and not always followed. Different agencies also have different ages for the transfer from children's to adult services – there sometimes appears to be a 'hole' between the two.

Access to information is a key issue. Most parents – and some professionals – are not aware of all the choices that may be available. Many students and parents have a narrow view of what is possible. They need to see what is on offer, i.e. be taken to visit. Many students and parents do not have the skills or resources to find information and make applications; they need support. Parents may feel intimidated in meetings with unfamiliar professionals where they may feel greatly outnumbered and excluded by their use of language and jargon. They may need access to support to enable them to attend meetings and express their views. There is a role here for advocacy, particularly to support those students whose families are not able to.

What are the major issues facing these young people and their families? These are the concerns most often raised:

- further education choices
- employment opportunities
- a home for the future
- benefits
- leisure and recreation opportunities
- relationships

Students with special needs have an entitlement to full-time education until the age of 19. Some students spend the last few years in an FE college. The transition from school to college needs to be carefully planned. Link courses are important for some students to facilitate a smooth transition. There is a need for a tightening of procedures in passing information about students with special needs from mainstream schools to colleges – some students may start inappropriate courses or do not get adequate support because the college had no information. What happens after 19 is often the crucial issue for students and parents.

Employment and day-care opportunities vary widely from area to area. Again just finding out what might be available can be a serious initiative test. Continuing education opportunities may be available, but again information is often difficult or impossible to find. Most parents and young people hope that there will be opportunities for employment. What is possible may depend very much on where you live. Increasing the number and range of supported employment schemes should be a priority. For those not able to work, finding appropriate daytime activities is a major concern. Although the situation is improving, the reality is that there is little or no choice and sometimes only part-time provision is available.

A place to live is another big issue. For the parents of those young people who will always need some support this is a very serious concern. Some young people remain in the family home from choice or by default, but ultimately the reality for most is that at some point they will have to move on when their parents are no longer able to care for them. It is now – belatedly – being acknowledged that young disabled people have the right to live independently, with appropriate support or care. Again the range and number of living opportunities vary widely from area to area. The reality for some parents is that the family reaches a point of crisis before the issue is addressed. Other parents may find the young person's need to live away from home is acknowledged, but no appropriate provision is available within the local area. Young people need access to a range of supported living options within the area to which they belong. The trend at present is to see living in small, family-sized groups in the community as the norm, but some young people may prefer to live in a larger group, while for others it may be their preference to live alone. Wherever they live, support needs to be appropriate and consistent. Work and leisure time need to be planned to meet the young person's needs, including opportunities for them to develop friendships and relationships.

The benefits system is another assault course. I cannot begin to explain its complexities – but believe me, it is complex, and mistakes are made. My advice, from personal experience, is, if in any doubt about entitlement, get expert advice. Clear information, accessible forms and a 'one stop shop' system are needed. Some young people with disabilities are now being offered the opportunity to choose and buy their own support. While this may seem a good option in theory, in practice this choice is only really accessible at present to those able to handle the money, tax, National Insurance etc., themselves. If direct payments are to be a realistic option for young people

with learning difficulties, the system needs to be rethought.

Accessing leisure and recreation opportunities is another issue that young people and their families face at this time. For some, as they get older, it becomes more difficult to be included in mainstream activities. Teenagers generally organise their own leisure – and often in ways we (as parents and professionals) would rather they didn't! There is a lack of appropriate, organised activities that include disabled young people. School and college holidays can be a particularly difficult time. The reality for some young people and their parents is a summer break with no organised activities, which can extend up to three months. For some parents who have managed to work, this is the point at which they are forced to give up.

The issues of relationships and personal safety can cause parents and young people much heart-searching. Schools and colleges may try to address these questions with students but staff often do not have appropriate skills and knowledge. This is an area where both parents and professionals need access to appropriate training. It can be a very difficult time for young people, who may be increasingly aware of their sexuality and need for independence – and for parents, many of whom find it very difficult to acknowledge that their very dependent child is becoming an adult.

Parents may struggle with other issues. They may feel a need to continue caring for their son or daughter, they may begin to worry about the future, when they are no longer able to care, or that might be too difficult to contemplate. Parents may recognise their young person's need for greater independence, but feel guilty about relinquishing the caring role. Some parents, usually mothers, who have been full-time carers, face a significant change in their lives if their son or daughter moves on from the family home. These parents need emotional, practical and sometimes financial support to move on themselves, to re-join the workforce and build an independent life. This may be the time when parents begin to agonise about who will look after their child's interests when they are no longer around, This may be a role for siblings, but what if this is not appropriate, or there are none? What about financial provision? Making a will to provide for the future of a young person with learning difficulties is another very complex issue.

To conclude here are a few of the highs and lows of my last few years as the parent of a son with Down syndrome 'in transition'.

- Eleven week summer holidays from FE College – with no access to organised holiday activity schemes.
- Hearing that he had secured funding for his place at residential college – after having been initially told by two professionals that he was not eligible to go.
- Receiving the cheque for £2,600 in unpaid benefits for my son – after one of the longest paper chases I remember.
- Days and days spent travelling the country visiting potential long-term placements for my son.
- Eventually finding the 'right' place – only to see the prospect of a place there disappear. My son, standing up and making a completely unprompted speech thanking, everyone for coming to his 18th birthday party.

Angelman Syndrome in the Media

Every now and again someone contacts us about articles that have appeared in the press, TV, and the radio.

Recently on Radio 4 Front Row there was a piece about Epilepsy in the Movies – which believe it or not has been the subject of research for some! One of the interesting observations was that Dopey, one of the seven dwarfs immortalised in the Disney Film, was a person with Angelman Syndrome. Walt Disney had been quite specific to the animators about how this character should be portrayed, leading to the supposition that he knew someone like this some 30 years or so before Dr Harry Angelman identified the condition. However, this has never been substantiated by Disney Studios. Prof Benard Dan and Florence Christiaens (Brussels Belgium) wrote a paper about this entitled "Dopey's Seizure" in 1999. The full article can be found on the Internet at www.idealibrary.com Many thanks to the Fox-Jones's who rang to tell us about this.

We also heard from Marks and Spencer in Harlow about one of our families who have been the beneficiary of their fundraising for the year – there was an article in the local paper and we were contacted by both Marks and Spencer and the local press. We hope that the money is helpful in supporting them to look after their son.

How ASSERT Can Help You

ASSERT is currently receiving an average of two or three phone calls a day from parents and professionals, and more recently enquiries via the website. The telephone also accepts faxes. All calls are going through to Sally Walburn. If you would like to speak to a particular trustee, Sally will be happy to put you in contact, or arrange for that trustee to call you back. Sometimes Sally is around but the answer-phone cuts in before she can answer and often people cut the call as soon as they hear the machine. If the number is withheld, obviously we are unable to call you back. So, please do leave a message!! In your reply it would be appreciated if you would let us know a time when it is best to contact you and also make sure your details are clear and not rushed, sometimes it can be difficult to decipher the number and name.

The support line and website enquiry form, is there for us to respond with suggestions and support, we cannot fight your battles but we may be able to put you in touch with someone else in a similar position. We can offer a listening ear and try to point you in the direction of further assistance. As with all support and advice, please remember that ASSERT is no substitute for professional assistance and if there are queries around medication etc., we are happy to talk to you but you must always seek the advice of your health professional. ASSERT will not endorse or support any one course of action but make suggestions for you to decide for yourself.

Josh Beds



All aboard the Night Express

Hi,
We thought readers of the ASSERT newsletter may be interested to hear about Josh Beds. Our son, Joshua, is six years old and has Angelman's UPD. Like many children with Angelman's, he suffers from sleep disturbances which can be fairly severe (for example, only a couple of hours' sleep a night at the worst point). When Joshua was small, he slept (or rather didn't sleep) in a cot. However, by the age of four, he was too big for a cot and so we had to look for an alternative. We quickly found that there was little on the market to cater for the needs of a child like Joshua. By this time, he was fairly mobile and we needed a bed that would keep him safe and secure. Our brief experiment with putting Josh in a standard bed turned out to be a disaster as we were woken at 3.00am to the sound of music from our living room (Josh had managed to turn both the TV and the stereo on) and nothing short of carnage in the kitchen, where he had raided the fridge and smeared the dog in coleslaw!

This left us with a problem, as we didn't want to put Josh in an outside cot. Instead, we wanted a bed that was safe and secure, like a cot, whilst also being good to look at and great fun for him.

We decided that as there was nothing on the market which met our needs we would design our own solution, and the first Josh Bed was born. As you can see from the photo, it is in the design of a train. It's a brightly coloured, fun piece of furniture

which is also very robust. Josh has been in this bed for a year now and it has withstood the most brutal of tests from our very strong and heavy handed little boy!

Josh loves his bed and we can lock him into it knowing that he will be safe and secure. He'll now happily lay in bed at night, wide awake watching a video snuggled up under his duvet rather than demanding our attention. This has been quite a breakthrough!

When we originally came up with the design, it was purely to meet Joshua's need. However, we have had such positive feedback from friends and Health professionals alike, that we have decided to manufacture them, providing the demand is there.

We should have a website up and running by the end of September

which will provide a lot more pictures and details. Our objective is to make quality and affordable beds that service children with disability. We will be producing designs for both girls and boys and are also in a position to meet individual design requests (within reason!). In conclusion, we want to provide beds that are fun, and not just functional, and which give dignity to our disabled children – in fact, furniture that any child would want to have in their bedroom.

If you are interested in knowing more, please do give us a ring on 01858 575144.



Off we go – into the Land of Nod – hopefully!

Andy & Fran Childerhouse

HELPLINE

**ASSERT Families/New Diagnosis/
Support Line**

01268 415940



ASSERT 2000 club

The ASSERT 2000 club was drawn at the Trustees meeting on 25th January 2004.

Winner G Meadows – £40

Second Ken Walburn – £20

Third L. Mason – £12

Congratulations to all. It's not too late to apply for the other three draws – cheques for £12 payable to ASSERT – please add your name and address and mark it for the 2000 club.

Development and behaviour in Angelman Syndrome:

– A study of development in 15q11q13 maternal microdeletions and behaviour across genotypes.

The above study was conducted by Dr. Angela Reason, Clinical Psychologist. This report is the second part of an overall study focusing on the **development and behaviour of people with Angelman syndrome**. Part one, already reported, looked at development in AS by deletion in individuals aged between 3 and 14 years. This second section focuses on behaviour across all ages and all genotypes.

Part Two: Behaviour in Angelman Syndrome

Background to the study:

Although a handful of studies exist looking at behaviour in Angelman syndrome (AS), the phenotype would still benefit from further exploration, using a wider sample including all genotypes and across the age span.

Aims of the study:

The aim of the present study was to:

1. Determine an inclusive profile of behaviour associated with AS, as reported by caregivers
2. Explore whether differences exist across gender, age and genotype

Procedure:

Recruitment took place via ASSERT. 76 families with AS children over 3 years old were sent a revised version of the **'Society for the Study of Behavioural Phenotypes-Postal Questionnaire'**. 43 returned the questionnaire, a response rate of 56.6%. Questionnaires were anonymous to ensure confidentiality.

Description of the sample:

Gender:	Males	67%	Females	33%
Age:	Children	83%	Adults (16 years +)	17%
Genotype:	Deletion	38%	Non-deletion	21%
			(all other groups)	

(Missing data 41%)

Results:

A general profile of behaviour was elicited covering the following areas: Feeding, Sleep, Social Behaviour, Language, Movement Activities, Unusual Movements/Interests, Self-injurious Behaviour/Aggression, Anxiety and Mood, and Skills. Space was also incorporated for comments from caregivers and analysed using a qualitative, more detailed approach. The main results for each area are presented below:

General profile of behaviour (whole sample)

Behaviour:

- One third of the sample had been referred to a clinician due to behaviour (unspecified); 14% had received medication as a result. Qualitatively, caregivers reported behaviours such as biting, pinching and hair pulling.

Feeding:

- One third ate 'more' or 'much more' than normal
- Over a third would attempt to access more food regardless of hunger
- Just under half were reported as faddy eaters
- 60% attempted to eat non-food items, which can present hazards
- 32% drunk less than normal amounts of fluid
- Qualitatively some behaviours associated with food included snatching and throwing, plus needing a routine around meal times.

Sleep:

- Approximately 60% reported their son/daughter did not have a regular sleep pattern and had difficulties settling off to sleep
- 81% reported their son/daughter woke up frequently during the night
- 86% did not report sleep occurring excessively in the daytime, indicating lack of sleep at night is not compensated for in the day.

Social behaviour:

- One third was considered to be 'isolated', 'in a world of their own', or 'unusual' in the way they use eye contact
- A high proportion were seen as friendly to strangers.

Difficulties with language:

(Due to restricted language in AS only one question from this section was used for analysis)

- 38% of parents reported their son/daughter had a tendency to scream or shout unexpectedly.

Movement activities:

- 64% of the sample were considered by their caregivers to be 'overactive'
- 75% were reported to 'create chaos aimlessly'
- 63% of AS individuals were reported to settle into activities they enjoy in 10 minutes or less
- 82% were described as clumsy or having problems with balance.

Unusual movements/interests:

- A large proportion of the sample (85%) were reported to display 'unusual movements'. Many caregivers noted 'hand flapping' as an example
- Just under half were thought to have unusual attachments to strange objects but there are likely to be differences in what people consider 'strange'
- Half the sample were reported to insist on routine and become upset with change
- 34% were thought to show unusual reactions to sound, light or smells.

Self-injurious behaviour/aggression:

- 95% stated behaviour did not reach a degree of violence that meant they required help
- Over half were reported to self-harm, but the question did not allow for context of this behaviour
- More individuals were reported to physically attack family members than non-family members, although this was not a problem for over half the sample. The increased amount of contact with family members may contribute to this difference, as well as primary caregivers being in a more disciplinary role
- 38% were reported to lash out but caregivers noted they felt this was not deliberately malicious; 30% felt their child deliberately destroyed items
- Stubbornness was considered a major problem in 33%.

Anxiety and mood:

- In keeping with the clinical feature of AS, 91% were not reported to be miserable or unhappy; 67% were seen as excessively happy.
- 81% reported an absence of serious temper tantrums
- Fearfulness was only reported in 26%
- 68% reported mood was often out of place (e.g. happy at sad occasions). This is likely to be associated with level of comprehension, and laughter as a feature of the syndrome.

Other:

- 60% reported their son/daughter to have unusually good skills (e.g. memory for people's faces and places, intuition for people's characters).

Help:

This was an open question asking caregivers to note areas in which they would like more help, which is important information

for service providers. The two main areas emerged as practical help (e.g. respite care, getting out socially, lifting and handling) and help with functioning (e.g. communication, self-help skills, mobility). More information on AS was also noted.

Differences in gender, age and genotype

Gender: Stubbornness was seen as a problem in more females, as was shouting/screaming unexpectedly. More females were reported to be 'isolated', 'in a world of their own' and display unusual facial expressions and gestures.

Age: More children than adults were found to attempt eating non-food items, have difficulty settling to sleep, and less likely to do nothing if left alone. This is likely to be developmentally relevant.

Genotype: More individuals of deletion type had a less regular sleep pattern and higher reports of unusual use of eye contact. Self-harm was higher in the non-deletion group, as was receipt of medication for behaviour.

Critical review

It is recognised that there are some problems with the measure used, such as undesirable use of language (e.g. 'normal', 'strange'), and asking parents to compare their son/daughter against other individuals of the same age.

Conclusions

In conclusion many of the behaviours profiled are more likely to be associated with level of development rather than specific to the AS phenotype. Secondly many of the behaviours noted are commonly observed in non-syndrome specific learning disabilities, such as hyperactivity and stereotypical movements. Areas in which the phenotype link appears stronger include sleep problems and interests in specific materials.

Exploring behaviours aims to create a more comprehensive knowledge of the phenotype which will aid diagnosis, provide a richer description for families, and enable treatment approaches to be more tailored.

I would like to thank all the families who took part in this survey.

If anyone would like to discuss issues relating to this study, Angela can be contacted on:

020 8921 3506 (direct line/voicemail)

or email: angela.reason@oxleas.nhs.uk

HELPLINE
ASSERT Families/New Diagnosis/Support Line
01268 415940

