

# Unique **XXYY** study day report

Families with a son with XYY – a single extra Y chromosome – met experts and researchers at a study day in Oxford, UK in April 2008. They heard what is already known from research conducted on boys and men with XYY who were identified in the 1960s and what is starting to emerge from a follow-up study of the development of children with an extra sex chromosome, launched last summer in the UK. This study will refine and update knowledge about boys and girls aged between 4 and 16 with Triple X, XYY and XXY (Klinefelter syndrome), and in particular what is known about their behaviour and learning.

## Emerging findings from the Sex Chromosome Trisomy study



**Gaia Scerif**, a lecturer in the Department of Experimental Psychology in Oxford, is a developmental psychologist interested in attention and behaviour and memory in the classroom. When she and Victoria

Leggett, the study researcher directly involved in meeting all parents, looked at everything published on boys who had been diagnosed with XYY, they found quite contradictory findings. Follow-up studies published around the same time on boys who had been diagnosed in the same way by screening every newborn baby had quite different findings. Why? They don't know, although one reason might be that the groups studied were small and the boys were tested in different ways.

The Sex Chromosome Trisomy study was designed to address these problems. There would be enough children studied – 100 boys with XYY, 50 diagnosed before birth and 50 diagnosed in babyhood or as children, as well as children with XXX and XXY. Where possible, a brother or sister of the same sex

“The boys have both great strengths and weaknesses.”

would act as a control, for comparison. The study would use standardised tests of language, cognition, behaviour, movement, communication and socialisation. They would aim to identify boys' strengths as well as their difficulties and they would compare the group diagnosed before birth with those diagnosed in childhood.

As a starting point the researchers have asked parents for as much information as they can give - on attention, communication, behaviour, movement, socialisation, dealing with the environment, the classroom and handling fears. They have asked about any impact of difficulties in the son on the mother's wellbeing.

Part-way through the study, what have they found? That these boys have both great strengths and weaknesses. On the one hand parents have said that their boys are caring, kind, creative, sporty, very good at computing and humorous. On the other hand they tend to have behavioural problems that are often due to frustration, difficulties with socialising and communication and difficulties at school.

### Attention and learning

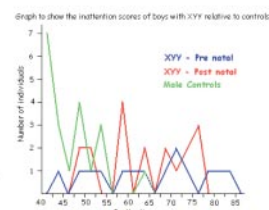
Gaia explained that everyone has an intuitive idea of what it means to pay attention. Yet attention actually has multiple components: it needs sustaining for a long period of time, it means not being distracted and being able to inhibit impulsivity. Most parents agreed that boys with XYY may have difficulty with paying

attention and controlling impulsive actions and said that this could create difficulties at home and in the classroom.

The researchers asked questions on attention and hyperactivity. Does your son seem to listen to what is being said or not? Does he have any difficulties in organising tasks and activities? Is he always on the go? One parent described their son's activity level as like having two packs of Duracell.

### Attention

- Significantly greater inattention
- True in all subscales measuring attention and hyperactivity
- Over 50% show high risk for ADHD
- No difference between pre/post natal diagnosis



Their findings show both how the boys do on average and also how they range – how different the children can be from each other. In terms of attention, the boys had significantly more difficulties than boys without XYY. These difficulties were true in all the subscales of inattention and hyperactivity. Up to 50 per cent were at high risk of ADHD (attention deficit hyperactivity disorder); a large proportion also have difficulties with impulsivity and for these issues there were no differences between boys who were prenatally and postnatally diagnosed.

A score above 70 on the graph would be in the range of clinically diagnosable ADHD. Most of the brothers without XYY (green line) scored around 50, so they had fewer problems. Among the prenatally diagnosed boys (blue line) there was a big spread, with one boy in

“Attention and hyperactivity seem to be weaknesses, although there is variability.”



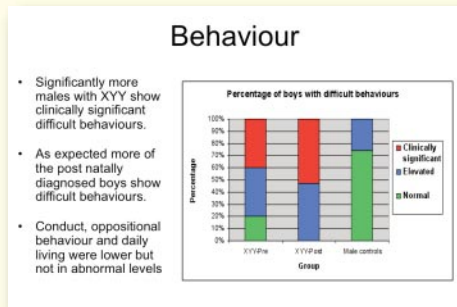


the 40s (few problems) and another in the 80s with a lot of inattention problems. The postnatally diagnosed boys tended to have more difficulties than their brothers but some were well within the normal range while others were in the clinical range. So prenatally and postnatally diagnosed boys had more inattention problems than boys who did not have an extra Y but there was a lot of variability.

“All the boys definitely want to be part of the group.”

## Behaviour

They asked questions about behaviour: Is your child impulsive? Does he act without thinking? Does he avoid social interactions? Does he have temper tantrums? Is he stubborn or sullen? Does he have a difficult time paying attention?

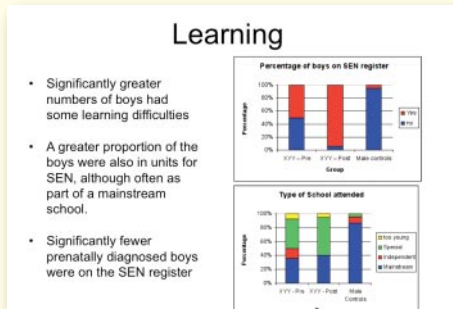


On average, they found that boys with an extra Y tended to show more clinically significant behaviours than boys without and this was more so for postnatally diagnosed boys.

## Learning

They asked what kind of learning provision the children need and what they have. One indication is how many of them are on the SEN register. They found that approximately half the prenatally diagnosed boys were on the SEN register and half were not. For the boys who were postnatally diagnosed the proportion on the SEN register was larger while only one of the brothers without XYY was on the SEN register.

What kind of schools do they attend? The



researchers found that a smaller proportion of the prenatally diagnosed group than the postnatally diagnosed group attend a special school or one with a special needs unit. Among the brothers without XYY only a small proportion are in a special school and most are in a mainstream school. Overall, around 40 per cent of both prenatally and postnatally diagnosed boys with XYY are in mainstream schools.

To sum up, the researchers found a mixed picture in terms of strengths and weaknesses. Attention and hyperactivity seem to be weaknesses, although there is variability, but this may impact on behaviour and may be reflected in the need for learning support.



**Victoria Leggett, a researcher at the Department of Experimental Psychology in Oxford, presented the team's findings on motor skills, communication and socialisation.**

## Motor skills

The researchers asked questions about both fine and gross motor skills such as: Can they fasten their buttons? Can they put a key in a lock and turn it OK? Can they manipulate small objects? How are they at running? Do they often trip over? Can they ride a bike?

Overall the boys did have more difficulties with both fine and gross movements than one would expect and there was no difference

“They are definitely good at a lot of things.”

between the prenatally and postnatally diagnosed boys.

How might this impact on communication and socialisation skills? For example, articulation might be quite important and might be linked to motor control. Victoria explained that by social skills we mean the ability to understand an interaction between two or more people and this can be understood on many levels. 'Firstly, if I point out something to you and am looking at you, that you have the same experience as I do. Secondly, that everybody doesn't have the same information so when your son with XYY comes home from school, whether he can tell you what he did all the way through or whether he misses out things that he did at school, thinking that you already know exactly what he knows.' She pointed out how difficult social rules can be - being able to queue, when to say goodbye, when to say please and thank you. Adults take social rules for granted but boys with XYY may need help with learning them. They may not read body language correctly or recognise emotions from people's expressions. They might find it hard to understand that others don't know what they do - so they step out into the road, thinking that the approaching car knows what they are about to do. They might not discriminate between people and use the same sort of language when talking to a teacher as they would when chatting to a friend. They might not notice if you're upset or getting bored because they have a difficulty with reading body language. One point with all the boys was that they do definitely want to be part of the group and are interested in talking to people but they may struggle to find the right way to join in.

## Communication

The most important thing in communication is

“If one considers whether they are considerate, kind and helpful or how much they think of others - they are.”

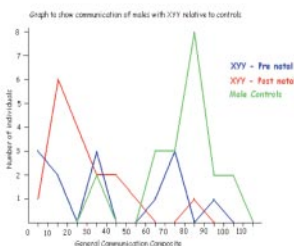


“Many of you have said the boys are happy, they're caring, they're sporty, they're creative and that they are really lovely boys.”

to convey what you mean. So the researchers asked: Do they understand the difference between cold (unfeeling) and cold (needs to put his coat on)? Do they pick up conversational overtures? Do they ask questions over and over even when they have the answer? Do they smile appropriately when talking to people?

## Communication

- Communication in boys with XYY is significantly worse than controls.
- No significant difference between pre and post natal diagnosis
- More of the pre natal fall within normal range.
- Prenatally diagnosed boys have preserved speech and syntax



Boys with XYY did struggle a bit more and on average there was no difference between prenatally and postnatally diagnosed boys. The majority of the controls score above 55 and a lot of the prenatally diagnosed boys also fall within that range and are doing fine. A lot more of the postnatally diagnosed boys are struggling more with communication and it's interesting that with the prenatally diagnosed boys a lot fewer needed speech therapy.

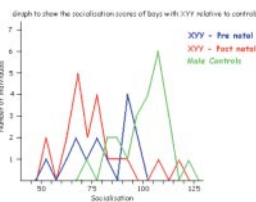
## Socialisation

They asked whether boys consider what others are interested in when talking to them. Whether they use words to express feelings like happiness or concern? Do they share things without being asked? Do they say sorry when they hurt others' feelings?

The boys with XYY did show poorer average socialisation than one would expect across all the subscales and there was no statistical significance between prenatally and postnatally diagnosed boys. Yet if one considers whether they are considerate, kind and helpful or how much they think of others – they do really well,

## Social Interactions

- Significantly poorer average socialisation
- True in all subscales measuring social interactions
- No difference between pre/post natal diagnosis
- Prosocial behaviour is a strength



which is important. It's easy to portray a negative picture and they are definitely good at a lot of things.

## Conclusion

In conclusion, the boys with XYY do score significantly different to sibling controls. But often their scores fall within the normal range. So they might score a little worse but usually it's within what one might expect within the general population. Difficulties with temper and difficult behaviours are often the result of frustration.

On a positive note, many parents have said the boys are happy, they're caring, they're sporty, they're creative and that they are really lovely boys.

“Difficulties with temper and difficult behaviours are often the result of frustration.”

## Hormones, Growth, Puberty and Fertility



**Gary Butler** is now Professor of Paediatrics and Growth at the University of Reading. He practises at the Royal Berkshire Hospital in Reading and at Great Ormond Street Hospital in London.

He explained the origins of his interest in children's growth. As a young paediatrician he had the opportunity to work with Dr Shirley Ratcliffe on following up the babies identified with an extra sex chromosome from 34,000 consecutive newborn babies screened over 13 years in Edinburgh. The study showed that chromosome variations are far from rare, affecting one person in 150, with sex chromosome variations affecting one in 250. So around 1:1,000 boys has an extra Y (XYY) and 1:1000 girls has Triple X (an extra X chromosome). Boys with an extra X chromosome are even more common at 1:700 or even less. But what difference does the extra sex chromosome make?

## Growth

At birth, boys with XYY in the Edinburgh group were the same size as XY boys (the controls). Growth in the early years continues fairly normally, but from the preschool years onwards most XYY boys are taller than average and a few are above the usual height range for their age. In terms of final height, the average for boys is about 176cm (5' 9-10"). The average height for boys with an extra Y chromosome is about 188cm (6' 2") so they gain about another 13cm-15cm (5"-6"). Throughout life children have lots of little growth spurts, with an extra spurt at puberty. Boys with an extra Y chromosome grow steadily faster throughout childhood and are usually already around 10cm (4") ahead before their puberty growth spurt, which comes slightly later.

What about body proportions? Weight is much the same; most of the height comes from more rapid growth in the legs, not in the spine or neck. The shoulders are the same width as in XY boys but the hips a bit wider. The head circumference is exactly the same. But these are subtle differences.

## Questions

**Q: When my son was diagnosed by amniocentesis the geneticist said that because we are both tall, he would reach 7 foot (2.13 metres), but an injection could be given behind the knee to control**





**growth. Our son is now 7 years old and already 4' 7" (1.40 metres). Recently a growth specialist said that our only option is to bring puberty forward. That seems like dabbling with nature but we are concerned whether our son will have the emotional maturity to deal with being so tall.**

**A:** It would be highly unusual to reach seven foot. We do sometimes give treatment to bring puberty forward a little sooner by giving natural amounts of the male hormone testosterone. This may reduce height by a couple of inches. The 'injection behind the knee' is an operation in which orthopaedic surgeons use a fine drill to take out the growing points above and below the knee. It's not particularly invasive but you would only do it when you don't want any more growth.

**Q: When do boys with XYY stop growing? About 18?**

**A:** A bit later than average. Most boys stop on average around 17 to 18, depending on the timing of puberty. A boy who goes through an early puberty will stop growing earlier and one who goes through a late puberty will stop later, whether he is XYY or not.

**Q: As a father, I'm very tall, but my wife isn't. Is he more likely to take after me?**

**A:** The link with family height remains. If your family is short, your son is likely to be taller than yourselves but if you are a tall family, he'll be that much taller. We usually assume that each parent has equal input but if one parent is taller than the other it depends whether that parent's genes are a stronger influence on the child or not and whether genes on the Y chromosome are controlling growth.

**Q: If a boy is already going through puberty and suddenly growing very rapidly, can you stop any further height gain?**

**A:** By the time they are going through puberty it's too late to use hormone treatment. If you want to restrict height, you have to take action earlier or opt for the knee operation.

**Q: My son doesn't seem to be particularly tall, although my husband is 6' 1" (1.85m)**

**and I'm fairly tall. But he is 11 and seems to be average in his class.**

**A:** He is likely to have a bigger than average growth spurt and end up above average. If he's average at 11 and has not yet gone through puberty he is unlikely to end up more than upper normal in height.

**Q: Is every boy with XYY taller than average?**

**A:** Not necessarily. The average XYY boy is 5" (13cm) taller than his father, one presumes because of the growth genes on the Y chromosome.

**Q: I've read that it's not hereditary but my husband's great-grandfather was 6' 8" (2.03m) and my daughter's quite worried.**

**A:** Apart from the genes on the Y chromosome, there are many other genes involved in height. Picture your own family and the spread of sizes even in your first-degree relatives and you can see what huge variations there are.

**Q: My son has had pain in his knees and last year in his hips and there was some concern that it might be Perthes disease. He was 6 last year.**

**A:** Hip problems can sometimes manifest as pains in the knees. If there's no evidence of inflammation and the joints are otherwise normal after a check-up, it may well be growing pains and simple painkillers are all that's needed.

**Q: During his many growing spurts, my son had a lot of cramp as well as growing pains. Is that typical?**

**A:** It may be. It's clear that there are lots of growth spurts throughout childhood and they are bigger in XYY boys. Growth spurts can hurt, especially when you've just gone off to sleep and your growth hormone switches on. But we don't know why they hurt!

**“In a crowd of 80,000 male football fans watching Chelsea play Manchester United last night, there would be 80 men with an extra Y chromosome.”**

## Puberty and fertility

Puberty is essentially normal. We rarely come across any problems associated with puberty in boys with an extra Y chromosome. Normally we see testicle development around 11¾, which is about six months later than in XY boys but well within the normal range. Pubic hair development is a little later than in XY boys, occurring on average at 12½ and in XYY boys at 13¾, but within the usual range. Testosterone levels before and during puberty are all entirely normal.

What about fertility? Looking at a Japanese study of 1000 men who presented to an infertility clinic, six per cent were found to have a chromosome variation but the commonest was Klinefelter's syndrome (XXY). Only a tiny proportion – 0.3% – had XYY and this could be due to other reasons than the chromosome variation. The simple message is that puberty and fertility are within the normal range.

## Questions

**Q: My son is only nine and we've noticed that he's started growing pubic hair. He's had tests that show that he isn't going through puberty yet. Is this normal?**

**A:** It may be; there are many possible reasons for getting pubic hair. It would be worthwhile seeing a paediatrician to get that checked out.

**Q: I was told by my health professional that puberty is very much delayed. How can we get the right message across and spread the XYY word?**

**A:** Doctors are supposed to practise on a scientific basis. The correct answer is that puberty in XYY boys is on average late normal.

## Learning

In terms of reading, one boy in six in the general population needs help while about half the XYY boys in the Edinburgh study needed help to get going with reading. As for mathematics there were no problems. At age 13, the boys with XYY were scoring better than the control boys. Standard IQ tests at 7 and 12 showed the average for XYY boys was over 100 and well within the normal range.

In terms of educational outcome, of the 17





boys who were followed up, two went to university and three went on to further education colleges. There was a wide range of employment: some ran businesses, some worked in service industries, others in the army or in community service, all very much what one would expect. The comment made by Dr Shirley Ratcliffe at the time was that there was some experience with multiple changes of job but no increase in the level of unemployment.

## Questions

**Q: In our experience, early diagnosis is very important not for medical reasons but for education. My son wasn't diagnosed until he was 14 when we asked for a chromosome test and the result is that he's missed most of his secondary education. He was constantly excluded from school because we and others weren't aware what was going on.**

**A:** Having a chromosome test is a difficult decision, because labelling can be both helpful and unhelpful. Our experience in the Edinburgh study is that the boys who needed extra help for reading were picked up by the schools who didn't know the diagnosis. Ideally that's what should happen – the educational process should be sufficiently sensitive to provide the support that children with difficulties need.

**Q: It was the social difficulties more than the educational difficulties.**

**A:** The problem then is that the resources for education are only enough for children with very severe problems who require a statement. Some children would benefit from extra help but the current funding can't provide it.

**Q: We found that our son didn't need a statement in primary school but he did in secondary school and we had a 2½ year fight to get one, so he lost a lot of ground.**

**A:** All is not lost. Learning can continue and if some ground is lost in the troubled adolescent years, there are continued maturation possibilities. Don't lose hope.

Professor Butler explained that neurological tests in the Edinburgh study had shown that some boys had slight intention tremors; others

had unusual tics and grimaces, but these were not particular to XYY and in themselves quite common. He said that in terms of co-ordination, the indicators pointed essentially towards normal development. He then turned to the tests of character and behaviour that were carried out on the Edinburgh boys with XYY.

## Character and behaviour

The Edinburgh study showed that the boys with XYY were slightly more prone than boys without an extra Y to physical concerns like stomach complaints and they had more speech problems. On the positive side, they had fewer eating problems and fewer sleeping problems. They were more likely to have a temper; they were a little more solitary; some boys had difficulty making friends; they were a bit more irritable; and some boys had a tendency to tell lies. In other things, the boys with XYY were no different from the controls, although minor things like stealing were a bit more common but as a group, they were much less fussy than boys with XY and had fewer worries.

About half the boys with XYY needed referral to a psychologist or psychiatrist compared with nine per cent of the control group. None of the boys had serious psychological problems – mainly defiant behaviour; temper tantrums, stealing and some wetting problems, all things you see in the population in general. Three boys did get depression but this can be environmental, you could find it anywhere. Why was the referral rate for boys with XYY so much higher than for boys with XY? In the XYY group, family problems were three times higher; mothers' depressive illnesses and marriage break-up was much commoner and it is likely that family issues were the principal reason for the possible behavioural issues.

## Questions

**Q: Is being attention-seeking part of XYY?**

**A:** My children – probably most children – are attention-seeking, so it isn't a specific part of XYY.

**Q: My son hasn't got many friends. He's an only child and his behaviour can be challenging. He can go up to children and**

**be in their face. I tell him to come back a bit and not frighten them. But it'll get worse as he gets taller.**

**A:** It may be possible to help with learning social adaptation skills.

**A (parent):** I've found that trying to get my son to join clubs or football teams or anything like that is helpful so as if he doesn't have a special friend, he does have groups of friends.

## Criminality

Professor Butler also mentioned the fourfold increase in conviction rates among the Edinburgh boys with XYY and the slightly lower age at conviction – 17 rather than 18 for the XY boys. But most of the crimes were minor and against property rather than persons and the most severe crime was committed by a control boy. The IQ of the boys with convictions was lower for both boys with XYY and for controls, so boys functioning less well intelligence wise were more likely to be led astray.

## Afterwards?

Quoting from Dr Shirley Ratcliffe, Professor Butler said in conclusion that the combination of slightly lowered intelligence, delayed speech development, emotional immaturity and greater height increased slightly the frequency of behaviour problems and criminal convictions. But there was improvement in time and spontaneous maturation and outcomes could be improved with consistent parental management and psychiatric help where necessary. So things can be done and help can be given.

## Questions

**Q: My son's nearly 13 and we've known for three years about the extra chromosome. Apart from when we were told about the extra chromosome, we've had no follow-up. Is that to be expected?**

**A:** It depends what the problem is. Having an extra Y chromosome is essentially a variation of normal. There is no particular health risk and the learning and behaviour variations are within what you see in boys who don't have an extra Y chromosome. If a boy's brought up in a





loving, secure, supportive home environment there's no reason why he should not do well. I tell my own patients that I'm there in case any problems arise but don't see them regularly as I'm not monitoring anything.

**Q: Does it make a difference to the outcome if the extra Y is in every cell in the body or not?**

**A:** Generally the chromosomes are tested on a blood sample, which is only one of the tissues in the body. You may see less of a marked effect generally in mosaic XYY [part XYY, part XY] but this is not always the case because it depends which part of the body has the extra Y. You are only checking a single blood sample but the brain or the knees might have more Ys.

## Males with an additional Y chromosome



**Pat Jacobs, Professor of Human Genetics at the Wessex Regional Genetics Laboratory, took families back to the 1960s and the early days of identifying and counting chromosomes.**

She showed how it was possible to

identify extra X chromosomes without a blood sample – just from a 'quick and dirty' method of wiping inside the cheek (buccal smear). Using these methods, a surprisingly large number of men with an extra X chromosome were found in the 'special hospitals' set up for people who are hard-to-manage but are not considered responsible for their antisocial behaviour. Even more surprisingly, many of these men in the special hospitals were found, on full chromosome analysis, to have an additional Y chromosome as well as the additional X – that is, their chromosome constitution was 48,XXYY. Men with this chromosome make-up are extremely rare in the population and to find relatively large numbers in the special hospitals had to be

significant. Professor Jacobs wondered if the additional Y was contributing to their reason for being in the special hospitals and, if so, would males with a 47,XYY constitution also be found in excess numbers in the special hospitals?

Professor Jacobs studied 315 men in the one special hospital in Scotland and found 9 men with XYY and one with XXYY. Eight of the 10 men were in the wing of the hospital for those with significant learning difficulties, described in those days as mental retardation.

At the time, the frequency of XYY in the population was not known and was only to be revealed by studies in six centres across the world where every newborn baby had their chromosomes tested. These studies covered 38,000 baby boys and found 43 with XYY, giving a frequency of 1 in 1,000. This showed then that there was a 32-fold increase over the population average in men with XYY in the special hospital in Scotland. But why were these men there? Later, careful clinical studies showed that the men were most likely to be in the group with a learning disability and that their crimes were much more often against property and much less often against people.

So ten males with an extra Y chromosome were found in the Scottish special hospital. But at that time in Scotland there were nearly two million men over 15 years old, so there had to be around 1,800 XYY men in Scotland. Where were the remaining 1,790? The answer is, Professor Jacobs said, 'We don't know. Are the XYYs in the special hospitals the tip of an iceberg or are the special hospital XYYs very atypical of XYYs in general?'

The researchers tried to find out. Looking at every prisoner in Scotland (889 males in all) they found one man with XYY. So the men with XYY were not in prisons. They looked in approved schools and found four, a small increase. 'But it is quite clear that the vast majority of adolescents and adults with XYY are not in the penal system,' Professor Jacobs stressed.

So where are they? 'What we need more than anything is to find out what boys and men with XYY are like,' she said. The only way to get an unbiased population is from a consecutive series of newborns. In theory, following up the

babies identified in the six centres across the world should have answered this question. But too many babies with XYY were not followed up – only 41 across all six surveys, so in each centre there was only a small number. Every centre followed them up in a different way, making comparisons difficult.

But Professor Jacobs said that babies recognised from prenatal diagnosis (usually undertaken because of the mother's age) can also be followed up. The Chromosome Abnormality Database, which records most chromosome abnormalities in Britain, has 192 records of baby boys born after XYY diagnosis. By following this group, the true picture of what boys and men with XYY are like can emerge.

Professor Jacobs stressed that most males with XYY are never referred for chromosome analysis. At her own laboratory in Wessex, covering the central part of southern England, just 16 per cent of all boys and men with XYY have been diagnosed. 'Is that because the remaining 84 per cent are perfectly normal? Is it because we don't know enough about them to know whether they should have been referred or not? I am delighted to hear that it would have been a huge comfort and help to the families here today to have known about the XYY diagnosis earlier.'

Lastly, Professor Jacobs gave some information on what people with an extra Y chromosome are known to die of; causes of death include health-related causes and accidents. She stressed again that her information only included those men with XYY who had been diagnosed – no-one knows about the 86 per cent or so who remain undiagnosed. The figures she showed also included a few men with more than one extra Y chromosome. The information demonstrated that 57 out of around 700 men with one or more extra Y chromosome had died, somewhat more than would be expected. There was no increase in deaths from cancer or from hormonal, metabolic, nutritional or mental disorders. There was, however, a rise in deaths from epilepsy – although with a total of four the numbers were very small – and also from respiratory and circulatory causes such as heart attacks and strokes. [Professor Jacobs later sent





additional information that showed that all deaths except four occurred in adults and 63 per cent of them – almost two-thirds – occurred in men aged over 50.]

“Pregnancies with XYY almost never seem to end in a pregnancy loss.”

## Family concerns

Families and teenagers broke into three groups to discuss issues around behaviour, social skills, learning, attention and concentration. These are the main issues that emerged.

### Younger group (primary school age children):

Families first discussed strategies for dealing with unacceptable behaviour, including a warning, followed by 'time out' on the staircase, with the time increasing by a minute with each extra year of age. One family reported that when their child was overwhelmed he was removed from the situation and given time to calm down and that he had developed his own coping strategy. At school children are given a cooling down time with a technique such as counting down from 10. One family pointed out that the way they treat their child now will affect his future behaviour; they noticed that their child can fly off the handle and can get stressed if he can't finish a task at the first attempt. Tantrums were reported to be instant, disappearing as instantly as they came.

Families then turned to early assessments of their child's special needs. They wondered: should they tell the school about the XYY diagnosis or not? Families with younger children were recommended to tell the school very soon so their son could have appropriate interventions.

Discussion then turned to when to tell the child. One boy recognised in himself his 'special thing', as he called his XYY. He was able to discuss with his mother why he was like this and found it helpful to know that he was not the only one. Humour was a great help in coping and families said they emphasised to their child that he is special, rather than different. Many families were told about

criminality at diagnosis and felt strongly that there was a need for better education, especially of medical professionals, about XYY.

Some families reported that their sons are easily led and over-friendly, talking to anyone including strangers. Many boys fixed on a specific person as a friend, even though that might be the 'worst' boy in the class. One or two parents said that they had to teach their child social skills as they didn't acquire them instinctively. Some children had unusual obsessions and others showed sexualised behaviour, which was not a problem at 10 but was if it continued beyond childhood.

In terms of learning, quite a few families said their sons fell into the autistic spectrum, some full blown, others with Asperger's. Several boys had speech and language difficulties and some were great fantasists. Some boys were very good at spelling in their heads but were not then able to write the word down. Some boys had no educational difficulties; others were more significant. Many boys functioned better in small groups. Many didn't like loud noises; others had a heightened sense of smell. One child hated having his hair cut.

Many boys were always hungry and constantly eating. Some were hyperactive, others complained of tummy aches but this might have been attention-seeking. Some were very good with mechanical devices. Many were not sensitive to the cold. In some, the message that they needed to go to the toilet arrived at the last minute. Some showed a fear of animals.

Reported by Beverly Searle, *Unique*

### Older group (secondary school age children):

Many of the concerns raised among the younger boys were also raised among the older boys. Behavioural issues were common, including an inability to control their temper particularly with objects, such as slamming doors. A number of boys were sensitive and cried easily. Families had tried many approaches to behaviour including taking fish oils and other supplements, consulting herbalists, behaviour therapy, star charts and time out. Some families had a positive experience with Ritalin (methylphenidate); it helped one boy to

organise his thoughts and Ritalin was very helpful with learning at school.

Many boys had slight obsessions: they were picky eaters; they didn't like foods to touch each other. Some had sensory issues, liking strong smells or soft clothes. Some had variable facial or vocal tics (clucking, grunting, tongue clicking) which were associated with stress. Some boys used a 'baby voice' or adopted different voices.

By secondary school age, most boys were so tall that people thought they were older than they were, creating inappropriate expectations. They were bullied more than others and this may have been associated with their height. A common feature was that they got tired easily and needed a lot of sleep.

In terms of learning, a number of boys had features on the autistic spectrum; some had a statement of special educational needs; generally a small class size, 1:1 help and homework support was helpful. Some found background noise distracting and managed better in a quiet environment. Some boys showed sexualised or inappropriate sexual behaviour or behaviour that might have been appropriate in a much younger child, in some cases attributed to a lack of social skills. Some boys tended to invade others' personal space. Families stressed that social skills training and repeating instructions for social skills was very helpful; role play also helped.

Families also discussed their sons' talents: some boys were extremely gentle and empathetic, some good with small children and animals (while others were scared of them). At school, many were brilliant at mathematics, especially mental arithmetic; one boy was very good at geography and people but was not interested in maths. As a group, they liked facts, science, puzzles, and construction toys like Lego; quite a few were creative, artistic and musical. They liked practical activities and computers. They were generally caring and got on well with their other brothers and sisters; they were very active.

A number of families reported improvements over time, often as leaps forward. Boys seemed to catch up at school once they had the right help.

Reported by Sarah Wynn, *Unique*





## Adolescents

Most of the discussion covered areas which other adolescents would be experiencing as well.

- All the boys except one were in mainstream school – one was in a special needs school having been expelled from a mainstream school for behavioural and anger issues.
- Likes – music (especially RnB and Heavy metal), video games (one boy showed almost an obsession), roller-coasters (all the boys showed a strong liking for very fast sports) and being outside rather than cooped up inside.
- Dislikes – school work
- They were quite happy with meeting new people, especially at events such as gigs or parties.
- They had normal relationships with their siblings – getting on at times and not getting on at times.
- School subjects – they found maths and ICT especially enjoyable and easy and were not particularly interested in languages.
- Sensory issues – high-pitched sounds were particularly annoying to them (eg the wireless system in the classroom). One boy didn't like the way cucumber smells and didn't eat it (though his family couldn't smell anything).
- Attention – most preferred writing answers down rather than having to speak up in class, though some were quite outspoken.

Reported by Satnam Juttla, *Unique*. The boys gave permission to report back the content of the discussion.

## Frequency of some features of XYY in *Unique* families

Feature	Number of boys (total 18)
■ Good visual memory	18
■ Mild mannered, lovely and caring	17
■ Fiery temper	16
– towards things rather than people	10
– towards people (in one case specifically family)	6
– partly or completely outgrown by the age of 15	3/3
■ Could be easily led	16
■ Stressed if they can't do something at first attempt	15
■ Best in small groups	15
■ Lack of danger awareness	14
■ Very active	14
■ Anxious at new situations, a change or anything different	14
■ Can be friendly to strangers	13
■ Autism formally diagnosed or possible autistic features	13
– formally diagnosed	6
– autistic features mentioned by professional staff	7
■ Heightened awareness of sound, inaudible to others or background	13
■ Any obsessions	12
■ Very easily tired – ever	12
– outgrown by 14 or 15 years	2/4
■ Good at practical things	12
■ Good at ICT	11
■ Better at maths than at language based skills	10
■ Not sensitive to cold	10
■ Clicks and tics under stress	10
■ Unusually strong	10
■ Indecisive	10
■ Attention-seeking	9
■ Fantasists – ever	8
– outgrown by 13 or 14 years	2/4
■ Heightened sense of smell	8
■ Artistic	7
■ Picky eaters	6
■ Fear of animals, including insects	6
■ Double-jointed	4
■ Under-challenged at school	3
■ Sexualised behaviour	3
■ Unusually weak	3

