

*Unique*

## Support and Information

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**www.aaa.dk/TURNER/ENGELSK/xyy.htm**

Online information about XYY from Denmark's Turner support group

Email support group at **http://groups.yahoo.com/group/xyy**  
Family advice and varied personal histories; not professionally moderated;  
represents families with problems, many of them unrelated  
to the extra Y chromosome.

*Unique mentions other organisations' message boards and websites to help families looking for information. This does not imply that we endorse their content or have any responsibility for it.*

This leaflet is not a substitute for personal medical advice. Families should consult a medically qualified clinician in all matters relating to genetic diagnosis, management and health. The information is believed to be the best available at the time of publication and has been verified by Professor Maj Hulten, Professor of Medical Genetics, University of Warwick, 2004 and by Professor Patricia Jacobs, Wessex Regional Genetics Laboratory, 2005.

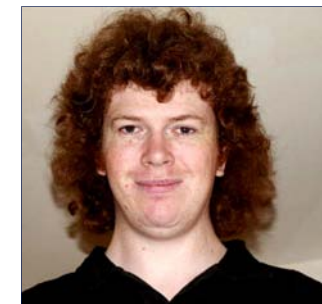
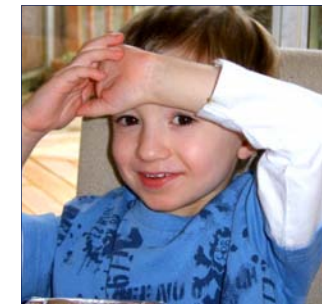
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*Unique*

**XYY**



## XYY

Read a leaflet like this and you could believe that boys with XYY chromosomes are a problem. For most families, this is not true. Overall, they are like any other child - a delight and a joy, but sometimes quite a challenge to bring up.

One boy in 1000 has XYY chromosomes. In the cells of their body, instead of 46 chromosomes including one X and one Y chromosome, they have one X and two Y chromosomes, making 47 in all.

Researchers who compared the number of boys found to have XYY because they exhibit problems with an estimate of the total number born concluded that the great majority of families, probably as many as 17 out of 20, never realise that their son carries an extra Y chromosome (Abramsky 1997).

A minority of families with a son with XYY chromosomes do encounter difficulties that test their parenting skills. This leaflet is written for these families, to share the experience and to provide some of the evidence they may need to access help.

**This is what some *Unique* families have said about their sons with XYY. These boys and young men belong to the minority who know that they have an extra Y chromosome.**

*“ Ted has a terrific sense of humour. He is very straightforward, with no ‘side’ to him - Ted, at 5*

*“ Ross is the kindest person I know. He would give you his last sweet and would take his brother’s punishment for being naughty without a second thought - Ross, at 13*

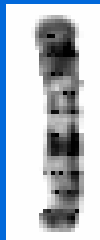
*“ Christopher is very caring, laid back and gentle, quite the opposite of an aggressive child - Christopher, at 13*

*“ Andrew is totally unmaterialistic. He has a great sense of righteous indignation, is appalled by suffering and angered by injustice to others - Andrew, at 17*

*“ Alistair has amazing resilience and an ability to keep bouncing back from adversity. He has a fascinating, somewhat off-centre mind and a child-like quality. His above average intelligence causes difficulty but is a strength as he can reason through difficult situations better than many people - Alistair, at 26*

*“ Tom can be extremely dignified in difficult situations - a genuine gentle giant - Tom, at 26*

*“ Most problems have improved or disappeared with age. Although life has been very difficult at times, especially in his childhood and teens, overall he is a loving, funny, interesting human being who has brought immeasurable joy and fulfilment to our lives - at 27.*



X chromosome

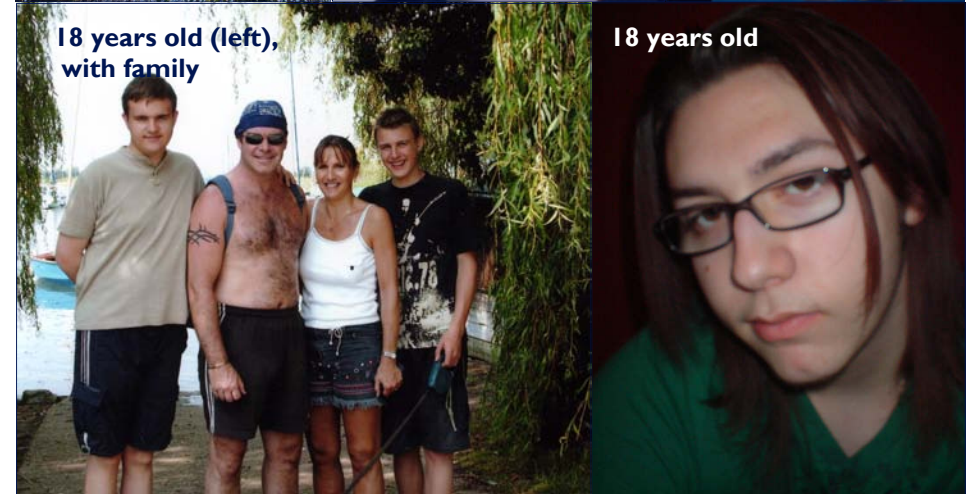


Y chromosomes

## Adolescents and men with XYY



21 years old



18 years old (left), with family

18 years old



36 years old



## Puberty

Unlike boys with XXY (Klinefelter syndrome), hormone levels in boys with XYY are generally similar to other boys and men.

There are no problems in boys with XYY associated with puberty (Linden 2002, 2; Linden 1996).

The Edinburgh study found that puberty started six months later than in XY boys but then proceeded normally. Pubic hair grew on average more than a year later in boys with XYY, around the end of the fourteenth year (Ratcliffe 1990; Stewart 1990).



## As adults

The Edinburgh study showed that most boys go on to play a useful part in their community and to hold down jobs as varied as a self employed businessman, a chef and a community service worker. Out of 19 boys, two studied engineering at university and three went to a technical college (Ratcliffe 1999).

The Danish study showed that one was majoring in science at college, while three others planned careers as a mechanic, engineer and farmer (Nielsen 1990).

Adult *Unique* members are working as a volunteer refurbishing computers for a third world charity, at a large supermarket, in agriculture and as a weapons expert. Those over the age of 25 generally live independently of their parents.



## As fathers

Men with XYY are as fertile as other men and do not run a risk of having children with a sex chromosome disorder that is discernibly higher than others. The extra Y chromosome is generally lost when sperm are formed, and although a tiny fraction of sperm cells with a double Y or an XY constitution do slip through, this can also happen to men with XY chromosomes. There is no real evidence either that men with XYY are any more likely to have children with other chromosome disorders than other men.

## Telling XYY boys

Most parents worry about when to tell their son about his extra chromosome. Not telling him leaves him prone to anxieties that may be much worse than the reality. The decision rests with the parents but once a boy with XYY becomes adult he has a right to know and even if the family does not tell him, professionals involved with his care may do so. One boy in the prospective studies discovered as a teenager when he read the correspondence from the clinic. It is important that the boy is informed together with someone who has wide experience of XYY and plenty of time and that, if he wishes, he can make contact with others of his own age with XYY.

## Telling others

There are two groups of people to be told: friends and family and school, doctors and any other therapists. It is the opinion of *Unique* that these people should be informed and given reliable supportive information on a strict need-to-know basis.

## Babies and young boys with XYY

“ This disorder is a difficult one to understand as they look very normal - parent



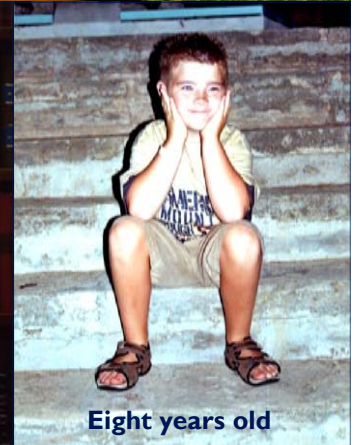
Four months old



Eleven months old



Six years old (left), with older brother



Eight years old



Nine years old



Nine years old



Ten years old

## Key features of XYY

- Normal appearance, often tall stature
- No increase in medical problems or illnesses
- Intellectual ability usually in the normal range, but an increased need for learning support
- No problems with sex organs or puberty
- Increased vulnerability to behaviour problems
- Increased vulnerability to stress

## Cause

Boys with XYY chromosomes have an additional Y chromosome from their father. In the great majority of cases, if not all, the two Y chromosomes failed to separate during the formation of sperm.

## Studies and references

What we know about boys with XYY chromosomes derives chiefly from seven prospective studies of babies whose chromosomes were examined at birth 25 to 35 years ago in Edinburgh, Boston, Denver, Winnipeg, Toronto, Denmark and Japan. Numbers of boys with XYY chromosomes in each study were small but the Edinburgh study is particularly useful because 20 boys were studied to the ages of 16 to 27. Almost all of these studies also examined 'control' boys as a comparison group, that is, boys who did not have XYY.

Now that increasing numbers of babies are being identified prenatally, researchers from Belgium have also compared a group of 12 boys with XYY chromosomes identified before birth with a group of 26 others who came to the attention of doctors as children or joined an XYY support association. A recent review of families' reports on 14 boys, all diagnosed during pregnancy, adds insights. Finally, a study of the psychology of 12 adult men not previously known to have XYY chromosomes completes the picture. This information is amplified by a survey of 43 members of *Unique* in 2003, referenced (U).

*Unique* families included boys diagnosed prenatally as well as boys diagnosed later. Families who are members of *Unique* and completed the survey are a self-selecting group, so the findings from this survey are likely to be biased towards showing an excess of problems.

The text contains references to the published studies, with first-named author and year of publication. This will allow you to search for the abstracts or original articles on the internet in PubMed or you can obtain abstracts and key articles from *Unique* (Geerts 2003; Linden 2002; Ratcliffe 1999, 1990; Maeda 1991; Nielsen 1991, 1990; Robinson 1991; Netley 1986; Evans 1990, 1986; Stewart 1990, 1986; Walzer 1990; Theilgaard 1986).



**One 10-year-old** is now succeeding at a small school where he is well known. As a young child, a quiet corner was made available to him when he felt unable to cope. At the age of 9, he received 10 hours a week of one-to-one support to help him manage his outbursts of anger.

**Christopher is a 13-year-old** with speech and language delay, a short concentration span and easy distractibility, lack of confidence and social skills. With unwavering parental backing and 12 hours a week of support at school, at age 13 in a mainstream school he achieved 10 excellent end-of-term grades, sixteen good grades and one satisfactory grade. Christopher does not have angry outbursts.

This advice comes from the mother of a **16-year-old** characterised as 'a very boisterous, energetic boy who can be affectionate but continues to suffer rapid mood swings': 'Be persistent. They wanted to put my son in schools for behavioural problems which I did not feel was the correct thing to do as he copies things and is easily led.'

## Alternative schooling

Four boys from *Unique* families (seven per cent) were being home schooled. Five boys (twelve per cent) have been expelled because of disruptive behaviour, the youngest at the age of five and one boy was expelled from every secondary school he attended. A 14-year-old who was only granted one hour of home tuition a day is now unable to read or write competently. Three boys have attended independent schools, two successfully and one with difficulties because allowance was not made for his behaviour.

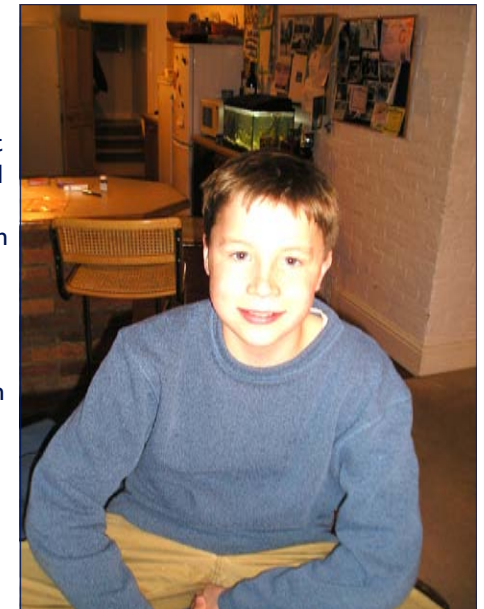
## Medical issues

Prospective studies suggest that boys with XYY have no particular medical complaints. The *Unique* survey confirms this but also suggests that among this highly selected group, seizures are more common than expected in the general population. Eight out of 32 family reports (25 per cent) mentioned seizures; this was by far the most common medical condition mentioned. In two children they occurred 'silently' as micronocturnal seizures. Two boys outgrew them in adolescence and there were no reports of seizures occurring in adults. Acne, by contrast, which has a known association with XYY (Robinson 1991), was mentioned by only four families (12 per cent).

## Teeth

Both deciduous and permanent teeth are somewhat larger in boys with XYY than other boys and the teeth have longer roots (Lahdesmaki 2004).

*"Glen is at a special boarding school where there is a philosophy of self advocacy and where social cognition is used to teach appropriate behaviour - Glen, at 17"*





The Denver researchers believed that the underlying problem was sensory motor dysfunction. 'In addition to affecting learning, the decreased perceptual organisation may contribute to diminished self concept and inadequate peer interaction,' they said (Salbenblatt 1987).

## Schooling

*“ Sean’s literacy and numeracy have improved since he left school and started work - Sean, at 27*

Boys with XYY usually attend a mainstream school, with or without support. A small number attend a special school. The choice of school type is to some extent subject to educational trends, so that all the boys in the Danish Arhus study were in mainstream schools while other studies showed a varying proportion of boys in special units (Geerts 2003; Linden 2002, 2; Ratcliffe 1999; Nielsen 1990).

## Statements of educational need

Combining the results of the *Unique* survey with information from the *Unique* database, forty-six out of 61 families (75 per cent) said that their sons had a statement of special educational need. Six boys (age 4, 5, 10, 13, 19 and 20) had a statement turned down. Most boys with a statement received one-to-one support for five to 25 hours a week. One young man had a statement throughout the school years but the request for it to continue at college was turned down. He found it very hard to cope and with hindsight his parents said that they would have appealed against the rejection.

### These two summaries illustrate differing needs of boys with XYY in schooling. Each is an excerpt from a statement of educational need.

*C’s language development and speech were delayed and he can be difficult to understand. His basic skills are very delayed. He has a short attention span. He has difficulty sustaining activities without adult prompting. He does not initiate interaction with others or spontaneously engage in classroom activities. He can be easily distracted. His impaired communications together with his passive nature have inhibited his development of age appropriate social skills ... he lacks confidence in new situations. He is very slow at writing - C, at 11*

*Recent assessments indicate that D’s learning potential is greater than most pupils with moderate learning difficulties. His behaviour is unpredictable and erratic. He can be attention seeking and verbally and physically abusive. ... is easily provoked, has difficulty sharing, turn taking and waiting for adult attention.... can become highly agitated and is impulsive and has difficulty coping with unfamiliar people and situations. D is reluctant to attend to items not of his own choosing, appears to be highly sensitive to sound and is distracted when presented with more than one stimulus ... he has difficulty controlling his anxiety and insecurities ... and is easily frustrated by his difficulties with learning and language ... he has difficulty with pronouns, prepositions and spatial relationships and can muddle his words and speak too quickly & loudly when excited or under pressure - D, at 14*

## Early intervention

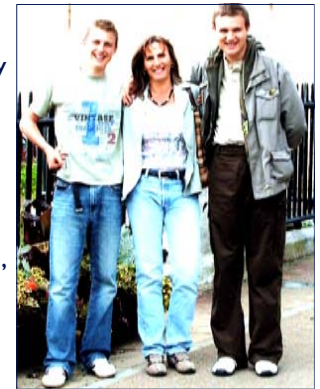
A number of parents document the successes achievable with early and continuing intervention.

## Height

Most men with XYY chromosomes are unusually tall. According to the Edinburgh study, XYY boys reach an average adult height of around 6’3” (188cm), on average 5” (13 cm) taller than other men. The *Unique* survey, a generation later, showed an even greater average adult height of 6’5” (196cm), with a range of 5’9” to 7’2” (175cm to 218cm). Heights of over seven foot (213 cm) were not uncommon.

There is some uncertainty about when the extra height (length in a baby) starts. The Edinburgh study showed that babies are not long at birth while the Belgian study showed that two out of three boys were extremely long at birth and 58 per cent had a birth weight at or over the 97th percentile. Whatever the birth weight and length, the Edinburgh study showed that growth speeds up from the age of two and by puberty a boy with XYY will be about three inches (7.6cm) taller than other boys. However, normal body proportions are retained (Geerts 2003; Ratcliffe 1990; 1986).

Among 26 families who told *Unique* when their son’s exceptional length or height was first noticed, eight remarked on it at birth or in babyhood, seven at pre-school, three in the primary school years and in eight boys the extra height was not noticed until puberty. Six out of 19 families with a son below 16 said that his height was not unusual and some boys were small for their age. One family with two six-foot sons remarked that the extra height in the boy with XYY was accounted for in leg length (U).



**Mother and sons: left, with XY; right, with XYY.**

## Being tall: advantages and disadvantages

Asked to suggest advantages and disadvantages of the extra height, parents suggested many more disadvantages. These included being picked on as a ringleader; a stooping posture; unfair expectations; size 17 feet; finding shoes and clothes; seating on public transport and bullying. The chief advantages were for sports such as basketball and as a boost for self esteem.

*“ I wasn’t aware of having XYY until a sudden growth spurt over a short period of time when I was in year 10/11. Being in an all boys’ school, I got a lot of verbal bullying, mainly name calling, which at times hurt, but I never let it bother me. After secondary school I moved on to art college where I found the students more mature. Over the past few years I have been attending yearly appointments with a doctor to monitor how tall I am and how much I weigh, to check blood pressure and give me the opportunity to ask any questions I have. My last appointment was this year and everything’s going okay. I wouldn’t change having XYY to be ‘normal’ because I like being as tall as I am and always getting noticed. Sometimes I get a bit paranoid in crowded places, especially when I hear laughter as I think it’s at my expense and nine times out of 10 I’m right, but I’ve grown up now, so I don’t care. It’s annoying having to duck under every doorway, but it’s become second nature. I try to be seen as normal and only realise how tall I am when I see my reflection next to a smaller person, or have photos taken with my girlfriend who’s 5’5” (165cm)*

*“ - James, 6’8” (203cm) at 18*

## Behaviour

Many boys with XYY pose parents with the ordinary challenges that all sons do. But the evidence from *Unique* suggests that around half of them exhibit styles of behaviour that make them stand out from the crowd. The others do not. Most studies and *Unique* agree that boys with XYY behave in certain characteristic ways, although this does not mean that the behaviour is necessarily caused by the extra chromosome. Formally, there is no precise behaviour type that is attributable to XYY, although a suggestion has been made that boys with XYY have 'extreme male' brains, making them less empathetic to others (affecting their social skills), decreasing their language abilities and endowing them with primarily analytical thinking (Geerts 2003).

All boys and all children thrive best in a home that is warm, stimulating and stable with parents who are alert to their needs and provide support and love. That is particularly true of boys with XYY.

### Behaviour: the evidence from studies

What do the prospective studies tell us? Almost half the boys in the Edinburgh study (47 per cent of boys with XYY against nine per cent of boys without) exhibited difficult behaviour (temper tantrums, defiance, stealing, bedwetting) severe enough to need an expert referral. The mothers of the Edinburgh study boys also reported more temper tantrums at home and personality questionnaires showed that the boys were 'more careless of social rules' (Ratcliffe 1990). The recent review of 14 boys diagnosed during pregnancy showed a varied behaviour pattern: seven boys were 'happy and outgoing', two were 'serious and quiet', three were 'moody'; five boys aged 8-10 had occasional temper tantrums and two boys of high school age had marked emotional problems. Overall four families sought psychological therapy (Linden 2002).

High drop-out rates may distort the findings of the other prospective studies but they do show a consistent range of behaviours. In Denver, the two boys remaining in the study after the age of 14 both showed a history of family conflict. One boy showed 'angry outbursts', the other remained subdued and resistant to schoolwork (Robinson 1991). In the Toronto study, two out of three boys showed severe temper outbursts when frustrated. One boy was shy, self-isolating and largely uncommunicative (Stewart 1990; 1986). At the age of 9, one boy had 'poor peer relations, marked irritability and temper outbursts' (Stewart 1986).



Temper tantrums and problems with concentration emerged in six out of 12 prenatally diagnosed boys in the Belgian survey. Among the 26 boys identified postnatally, the range of behaviour difficulty was wider: shyness (in 73 per cent), aloofness, aggressiveness and impulsive behaviour and petty crimes were recorded. In four, a diagnosis of pervasive developmental disorder was reached – 100 times the rate in the general population (Geerts 2003). Pervasive developmental disorders are a group of disorders characterised by delays in the development of socialisation and communication skills.

such as pronouns and prepositions appear to be common language problems, leading to hesitancy and difficulties in conversation. Speech problems are especially noticeable in social situations. A minority develop a stammer.

The *Unique* survey confirms that speech and language problems are not inevitable. Even among this highly self-selected group, the two oldest *Unique* members, in their twenties and thirties, report no problems. On the contrary, one has 'an excellent vocabulary and above average communication skills'.

### Interventions

The *Unique* survey showed that where speech therapy was offered early, it prevented problems in the primary school years and long-standing speech difficulties were less likely to occur. Some boys need speech therapy throughout childhood.

### Affecting the ability to learn: attention and concentration

#### Evidence

The prospective studies show that easy distractibility and hyperactivity occur more frequently among boys with XYY than XY boys (Evans 1990; Walzer 1990). The eleven boys in the Boston study were often off-task or obviously anxious during assessments. Six of the 11 needed support for attention problems and in most, this became evident around the age of 2½ or 3. The boys' primary school teachers frequently reported restlessness or running about when they were expected to be quiet (Walzer 1990).

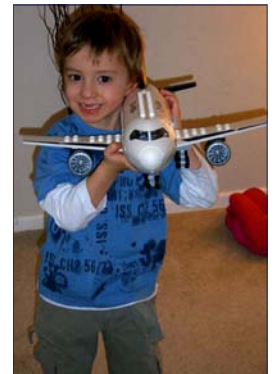
Similarly, postnatally diagnosed boys in the Belgian study had rates of 27 per cent for concentration problems (7.5 per cent in the general population) and 11 per cent for attention deficit hyperactivity disorder (ADHD) (Geerts 2003). The rate in the Denver prenatally diagnosed boys was 21 per cent (Linden 2002).

Many families in the *Unique* survey mentioned poor concentration as a cause for their son's education difficulties. Sixteen boys (33 per cent) were described as hyperactive or had received a diagnosis of attention deficit disorder. The youngest child to receive a diagnosis was 3, although the mother of a young man of 34 commented that she was aware of her son's hyperactivity by twelve months but could not persuade doctors to agree with her. A small number of boys were on prescribed medication, specifically methylphenidate and a recent study has underscored the importance of giving boys with XYY who have attention and concentration disorders a full neuropsychological evaluation and where appropriate a trial of stimulant medication (Ruud 2005).

### Affecting the ability to learn: social skills

Fifty-eight per cent of *Unique* families noted a lack of social skills. The components of social difficulties are complex, but include difficulties with turn taking and negotiating, ignoring social cues, inability to see other people's points of view, tactlessness and anxiety in groups.

The parents of three adult men with XYY commented that their sons found group situations especially hard to handle and were therefore disadvantaged at school. Once they left school, their difficulties became less obvious.



with six per cent in the general population (Geerts 2003; U).

In the Edinburgh study two boys had intention tremor – a tremor that is worse during voluntary movement - and two had facial tics (Ratcliffe 1999). Other studies confirm these findings, showing intention tremor as both more frequent and more intense in XYY (Theilgaard 1986). The tremor is reported in 4/26 boys in the Belgian group of boys diagnosed postnatally (Geerts 2003).

### Interventions

Importantly for prevention and early intervention strategies, three of the four boys with the lowest neurological or sensory motor integration scores in the Denver study walked slightly late, at the age of 14 to 16 months. The researchers imply that late walking could be used as an early marker for boys with XYY who would benefit from occupational and physiotherapy to improve co-ordination. They recommend that all boys should be reviewed regularly and intervention started as early as possible (Salbenblatt 1987). Evidence that this is not happening comes from the *Unique* survey which shows that despite very high rates of co-ordination and balance problems, only just over half of the *Unique* boys (13/25) aged under 19 received any occupational therapy, while fewer than half (10) received physiotherapy.

“ *Asher has an incredible visual memory. He can spell anything he has seen once but has huge difficulty with writing and if possible won't do it* - Asher, at 10

“ *Alistair writes competently but has poor hand co-ordination so it looks untidy; he uses a computer to express himself* - Alistair, at 27

“ *Matthew has excellent creative writing skills and excellent keyboard skills but unformed writing* - Matthew, at 34

### Affecting the ability to learn: speech delay

While most boys start to talk when you would expect, some are slow to speak. With or without speech therapy, most boys overcome their difficulties by teen age.

### Evidence

The studies indicate speech delay in up to half of all XYY boys. The Edinburgh study showed that 42 per cent of XYY boys had speech delay compared with 18 per cent of XY boys (Ratcliffe 1999). The Denver study (just four boys) showed that all had mild language delay, speaking two-word sentences later than 28 months. Follow-up at age 6-9 years showed that two boys still had a slight delay while two had a moderate language impairment (Robinson 1991). Similarly, half of the 12 boys identified on amniocentesis in the Belgian survey experienced speech delay and two of the 14 prenatally diagnosed Denver boys had speech therapy (Geerts 2003).

Evidence from *Unique* shows that speech delay can be obvious as early as 18 months. There is a wide range of severity, with some boys only experiencing mild difficulties in the pre-school years. In addition to problems with expression and with articulation (the typical speech style is described as 'slushy'), some children show delay in understanding. A difficulty in summoning words or the correct word and omission of parts of speech

Early studies in a medical-penal setting, that is, special hospitals, showed that over three per cent of men there had an XYY constitution. As only 0.1 per cent of newborn boys have an XYY karyotype, this represents a 30-fold increase of such men in special hospitals (Jacobs et al 1965). There was a strong link with learning difficulties, so that the rate of men with XYY and a learning difficulty was over four percent, while it was less than half that among men with no learning difficulties. These findings show that a small proportion of XYY boys and men have a marked level of anti-social behaviour. However, while this is substantially increased over the level in XY men, the vast majority of XYY men and boys do not show this extreme level of anti-social behaviour. These studies are now almost two generations out of date and there is an urgent need for a behavioural study of XYY males who were diagnosed before birth.

### Behaviour: the evidence from *Unique*

“ *It was as if we were dealing with a teenager for 15 years.* The *Unique* survey largely confirms these findings. It shows that **temper tantrums** almost certainly occur independently of frustration at not being able to communicate easily, although they may be triggered by it. Bouts of aggression occur in otherwise perfectly happy, loving boys. They can start as early as 15 months,

but more typically develop around age 5 to 6 and in some do not occur until the teen years. Triggers for tantrums include children being denied what they want and social stress, including stress at school, tiredness and unexpected changes in routine. Behaviour during a toddler tantrum can include throwing objects including furniture, hitting people (including strangers in the street), headbutting, biting and spitting. As boys get older, the tantrums may become more openly aggressive or violent, and behaviour includes swearing, shouting, screaming, kicking and pinching as well as offensive rudeness.

**Emotional immaturity** and **impulsiveness**, exacerbated by the disparity between apparent and actual age, was noted by half of the *Unique* families. This was also confirmed in 8/14 boys aged 7-16 and in XYY men by an in-depth psychological study (Theilgaard 1986; Linden 2002, 2). Twenty-three of the survey families (53 per cent) confirmed that **lack of confidence** was a problem in childhood and in the teen years. Other behavioural traits are mentioned much less frequently. Some boys show **obsessive** features. A minority record stress-related conditions.

### Handling behaviour: families' advice

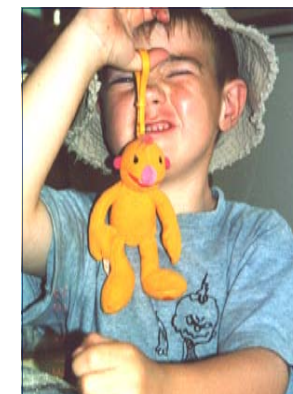
Age 26: Never give up

Age 22: Seek help as early as possible.

Age 19: Keep a simple set of rules and do not allow straying from them without an immediate consequence. But give praise at every conceivable moment. Avoid pressure, don't ask too much or too little and take one step at a time.

Age 14: Be there for him until he realises that he is the only one who can help himself.

*Unique* families report greatest success when they can be patient, calm and firm in their approach. Positive parenting techniques help, with predictable routines and clear





boundaries and expectations. Talking problems through and encouraging the boy to talk is helpful, especially when problems are due to emotional immaturity. The home atmosphere needs to be 'firm but flexible' and boundaries need to be clearly set. Boys up to age 11 may respond to rewards and star charts and by secondary school age boys may be motivated to help themselves. But many parents admit that despite their best intentions, the best support and the most structured home environments, the answer was professional help and medication. Both the studies and *Unique's* experience showed that behavioural treatment was hard work but successful. One family described medication (accepted reluctantly) as 'an oasis'. Counselling and prescribed medication as well as natural maturity improved the behaviour of the Edinburgh boys (Ratcliffe 1990).

## Mood disorders

### Evidence

There is no formal evidence of an increased risk for schizophrenia or psychiatric disorders (Mors 2001). The evidence from the prospective studies is that a minority of boys have a heightened negative response to stress. The Edinburgh boys were more anxious and three had a depressive stress reaction (Ratcliffe 1990; Netley 1986). One boy in the Denver study showed a high rate of anxiety due to unexpressed fears and worries (Robinson 1991) and the Winnipeg boys scored higher than their brothers and sisters on depression and withdrawal (Evans 1986). One teenager in the Denver review of boys diagnosed in pregnancy also developed depression and mood swings (Linden 2002, 2). The rate of mood and psychiatric disorders among *Unique* members is unknown, but 11 individual reports from 36 families of boys with XYY aged over 15 show severe mood swings in four, depression or bipolar disorder in six, panic attacks in one and schizophrenia in one. One family in the Denver review of boys diagnosed in pregnancy recorded bipolar disorder in their adolescent boy with XYY.

### Interventions

Many *Unique* families were offered psychological or psychiatric help or help with social skills. In some cases parents saw psychiatrists separately to learn how to manage their son's behaviour, but with increasing age and certainly from puberty, behavioural intervention or psychiatric help was offered direct to the boy.

No single programme answers all the needs of a family affected by XYY. Three themes emerge from the *Unique* survey:

- families need consistent, imaginative and proactive help. They value interventions that develop skills within a framework shared by other children without a chromosome disorder
- boys develop best in less stressed families. Having a son with behaviour or mood problems is intrinsically stressful and support services need to direct interventions to the family as well as to the boy with XYY
- support should continue beyond adolescence into adult life.

*“ There has been no help anywhere for Jack as he has got older. Sometimes he needed to be able to talk to somebody other than his parents, but this isn't classed as an illness or a problem, so there is no one to refer to - Jack, at 19*

## Learning

Boys with XYY chromosomes have a tendency to minor educational difficulties and in general, twice as many boys with XYY as boys with XY chromosomes need learning support, particularly with literacy in primary school.

The combined results of the large prospective studies showed that most boys' IQ is within the normal range, averaging 105 points and ranging from 65 points to 129 (Ratcliffe 1999; Stewart 1990). Compared with their brothers, boys showed a loss of 10 to 15 points, no more than the difference that might be found between brothers and sisters in any family. The recent Denver review found one boy who was outperforming his brothers at school. It also showed above average general performance, with 12 out of 14 boys achieving A and B grades and two boys receiving C and D grades. Boys generally did best at mathematics, science and reading and eleven boys were especially good at science.

Researchers agree that many boys need extra learning support. In the Denver study, all boys needed extra help with reading, language or writing (Robinson 1991). The Boston study showed that XYY boys frequently presented management problems for the teacher. However, their academic histories were variable and 4/11 were planning to attend college (Walzer 1990). In Denmark, 3/6 boys needed remedial teaching (Nielsen 1990). Despite their high academic grades, five of the 14 prenatally diagnosed Denver boys had extra learning support and two were diagnosed with learning disabilities (Linden 2002, 2). Specifically, while showing no difference in mathematical skills, over half (54 per cent) of the Edinburgh boys had difficulty reading, compared with only 18 per cent of other boys.

### Affecting the ability to learn: fine motor control

#### Evidence

When four boys of primary school age identified in the prospective studies were compared with boys with XY chromosomes, the XYY boys had mild to moderate problems with sensory-motor integration, motor planning, co-ordination of both sides of their body, hand-eye coordination and decreased muscle tone. To complete tasks, they needed extra multi-sensory clues. Their finger joints tended to be hypermobile and they had a poor grip, hindering writing skills. 'The neuromuscular status of the 47,XYY boys was poor and remarkably consistent,' said the Denver researchers. This contributed to school performance below that expected from their intellectual potential (Salbenblatt 1987). Among the Denver boys diagnosed during pregnancy, five/14 were described by their parents as being poorly co-ordinated (Linden 2002, 2). The Edinburgh study also revealed a slight deficit in fine motor control, meaning that boys were more likely to face problems with tasks such as writing, drawing and cutting with scissors.

These observations are amply supported by the *Unique* survey, in which 36 families (84 per cent) recorded a problem with either fine motor control or balance and by the Belgian study in which one in four boys had delayed motor development compared

*“ Top of his class in maths, no problems with memory but his reading is one year behind his age group*

