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Review Article

A REVIEW ON JACOB SYNDROME

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XYY syndrome is a rare chromosomal disorder that affects males. It is caused by the presence of an extra Y chromosome. Males normally have one X and one Y chromosome. However, individuals with this syndrome have one X and two Y chromosomes. Affected individuals are usually very tall. Many experience severe acne during adolescence. Additional symptoms may include learning disabilities and behavioral problems such as impulsivity. Intelligence is usually in the normal range, although IQ is on average 10-15 points lower than siblings. In the past, there were many misconceptions about this disease. It was sometimes called the super-male disease because men with this syndrome were thought to be overly-aggressive and lacking in empathy. Recent studies have shown that this is not the case. Although individuals with XYY syndrome have an increased risk for learning disabilities and behavioral problems, they are not overly aggressive, nor are they at an increased risk of any serious mental illness. Because these boys are at a higher risk for having learning disabilities, they may benefit from speech therapy, tutoring, and general awareness of the specific issues they struggle with. Although the first years of school may be more challenging for boys with XYY syndrome, they generally go on to lead full, healthy, and normal lives.

Keywords: Jacob syndrome, XYY, Karyotype, Criminal convictions, Impulsiveness.

INTRODUCTION

Rare Chromosomal Disorders

The term, 'rare chromosome disorders', refers to conditions which:

1. Occur due to missing, duplicated or re-arranged chromosome material.
2. Have a low prevalence rate (thus not including chromosomal disorders such as Down syndrome). Chromosomes are structures found in the nuclei of cells in human bodies. Each chromosome contains thousands of genes which determine how we grow and develop. A typically developing person will have 23 pairs of chromosomes with one member of each pair being inherited from each parent, giving a total of 46 individual chromosomes. Two of these are the sex chromosomes, which determine whether we are

female (XX) or male (XY). The remaining 44 chromosomes are grouped in 22 pairs numbered.[1]

TYPES OF RARE CHROMOSOME DISORDER

1. **Rearranged abnormality:**
Students with a rearranged abnormality may not experience any symptoms, but might have problems in reproduction. Some people consider genetic counseling when they are planning a family to assess the potential impact their chromosomal abnormality may have on their children.
2. **Missing or duplicated chromosome material:**
For students with missing or duplicated chromosome material, the effects will vary, but symptoms could include physical and/or health problems, learning disability and maybe challenging

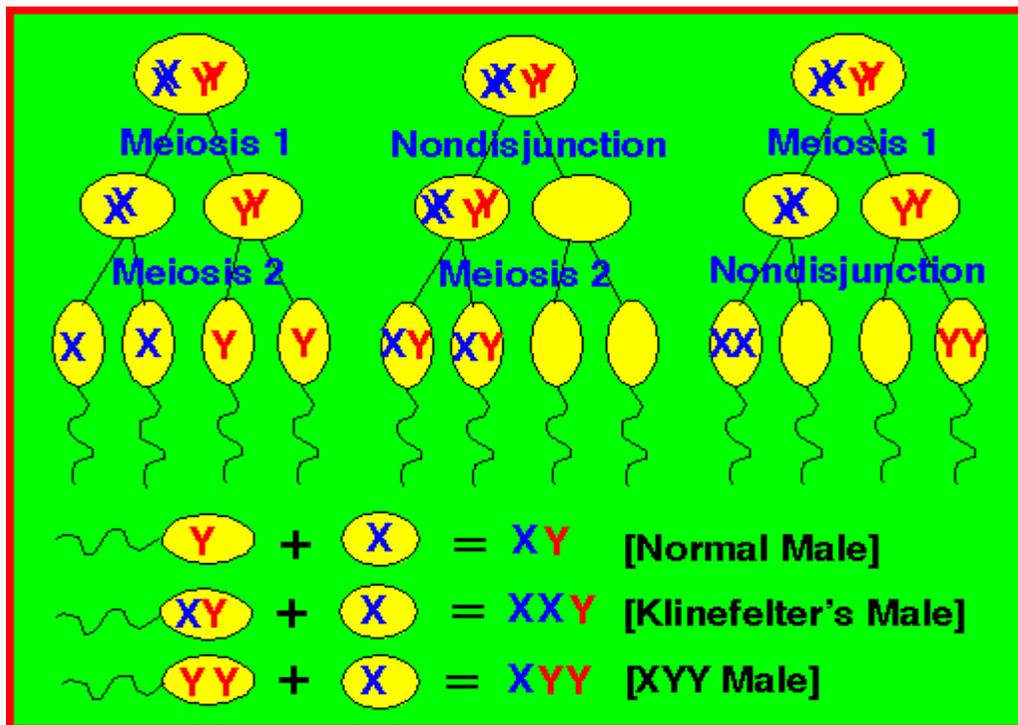


Fig. 1: Post fertilization phase

behavior .The combination and severity of symptoms will vary depending on which sections of chromosomes are involved, and the ways in which they are different.

3. Chromosome loss or gain:

Students with a loss or gain of chromosomes will experience some degree of learning disability and developmental delay. This is thought to be more serious than the presence of an extra copy of the same part of an extra copy of the same part.

Chromosome

The part of a cell that contains the genes which control how an animal or plant grows and what it becomes threadlike DNA-containing structures of cellular organisms that are located in the nucleus of eukaryotes, are usually ring-shaped in prokaryotes as bacteria), and contain all or most of the genes of

the organism.[2]

Chromosomal abnormalities

The prevalence of chromosomal disorders cannot be fully and accurately determined because many of these disorders do not permit full embryonic and fetal development and therefore end in spontaneous abortion. About one in every 100 newborn infants do ,



Fig-2: Chromosome however, have a gross demonstrable chromosomal



abnormality. A large majority of cytogenetic abnormalities can be identified by cytogenetic analysis either before birth, by means of chorionic villus sampling or amniocentesis.

Cytogenetic disorders with visible chromosomal abnormalities are evidenced by either an abnormal number of chromosomes or some alteration in the structure of one or more chromosomes. In the language of the geneticist, trisomy refers to the presence of an additional chromosome that is homologous with one of the existing pairs so that that particular chromosome is present in triplicate. An example of this type of disorder, which produces severe anatomical malformations and profound mental retardation.

Gene

A part of the DNA in a cell that controls the physical development, behaviour, etc. of an individual plant or animal and is passed on from its parents: The illness is believed to be caused by a defective gene. The scientists identified a defective gene. A particular gene is responsible for the inheritance of eye color. This mutant gene is thought to cause cancer. It is impossible to say how much a person's behavior is predetermined by their genes. Our personalities result from the complex interplay between our genes and our environment.

Genetic inheritance

Genetic inheritance by which a dominant gene is carried on the X chromosome. As an inheritance pattern, it is less common than the X-linked recessive type. In medicine, X-linked dominant inheritance indicates that a gene responsible for a genetic

disorder is located on the X chromosome, and only one copy of the allele is sufficient to cause the disorder when inherited from a parent who has the disorder.

X-linked dominant traits do not necessarily affect males more than females (unlike X-linked recessive traits). The exact pattern of inheritance varies, depending on whether the father or the mother has the trait of interest. All daughters of an affected father will also be affected but none of his sons will be affected (unless the mother is also affected). In addition, the mother of an affected son is also affected (but not necessarily the other way round). Some scholars have suggested discontinuing the terms dominant and recessive when referring to X-linked inheritance due to the multiple mechanisms that can result in the expression of X-linked traits in females, which include cell autonomous expression, skewed X-inactivation, clonal expansion, and somatic mosaics.[3]

Sex-linked Disease

Sex-linked diseases are passed down through families through one of the X or Y chromosomes. X and Y are sex chromosomes.

Dominant inheritance occurs when an abnormal gene from one parent causes disease even though the matching gene from the other parent is normal. The abnormal gene dominates.

But in recessive inheritance, both matching genes must be abnormal to cause disease. If only one gene in the pair is abnormal, the disease does not occur or it is mild. Someone who has one abnormal gene (but no symptoms) is called a carrier. Carriers can pass



abnormal genes to their children. The term "sex-linked recessive" usually refers to X-linked recessive difficulty may be press Possible indicators of rare chromosome disorders.

JACOB SYNDROME

Other names:

- Jacob's syndrome
- XYY karyotype

XYY Syndrome

Jacobs's syndrome: Rare Disease Status

Jacob's syndrome is listed as a "rare disease" by the Office of Rare Diseases (ORD).

- Synonyms of XYY Syndrome
- 47, XYY syndrome
- Jacob's syndrome
- XYY karyotype
- YY syndrome

History

In July 1999, Psychological Medicine published a case-control study by Royal Edinburgh Hospital psychiatrist Michael Got and colleagues that found an increased rate of criminal convictions among seventeen XYY men identified in the Edinburgh newborn screening study compared to an above-average-IQ control group of sixty XY men, which multiple logistic regression analysis indicated was mediated mainly through lowered intelligence.[4]

In June 2002, the American Journal of Medical Genetics published results from a longitudinal prospective cohort Denver Family Development Study led by pediatrician and geneticist Arthur Robinson, which found that in fourteen prenatally diagnosed

47,XYY boys (from high socioeconomic status families), IQ scores available for six boys ranged from 100–147 with a mean of 120. For the eleven of fourteen boys with siblings, in nine instances their siblings were stronger academically, but in one case the subject was performing equal to, and in another case superior to, his siblings.

XYY individuals among the 197 males tested-an incidence of 3.5%. Subsequent examination of these eight XYY individuals revealed no physical abnormalities except a tendency to be taller than the other patients.

Assessments of the behavioral characteristics of these individuals, on the other hand, showed that although none of them suffered from a true psychosis, each suffered from a severe, indeterminately caused personality disorder. [5]

They were "unstable and immature, unable to conduct adequate personal relationships, showing a tendency to abscond from institutions and committing apparently motiveless crimes, mostly against property."

A similar study, conducted concurrently by another research group, produced consistent findings."° Again, a high percentage of XYY individuals was found among antisocial or criminal types as compared to a low incidence discovered among mentally diseased and normal individuals. In addition to these studies newspapers around the world began reporting the lurid crimes of a few men with an extra Y chromosome.

The XYY syndrome was first offered as evidence in the Paris murder trial of Daniel Hugon. Hugon's



attorneys revealed that his sex chromosome structure was XYY rather than the normal XY and argued that this anomaly caused his violent behavior. [6]

Speculation then began to grow in this country that Richard Speck, convicted killer of eight nurses in Chicago, was also an XYY and that this fact might affect his pending appeal. 6" Speck's prominence catapulted the XYY syndrome into a storm of controversy which exploded with the acquittal by reason of insanity of an XYY defendant. Over the last two decades considerable efforts have been made to further refine XYY studies and the effects of the syndrome on behavior. The early studies are now considered to be significantly biased. Much of the information for these studies was obtained through screenings of populations selected because they demonstrated a particular personality trait or physical characteristic.

The studies undertaken to search forth XYY condition were characterized by efforts to identify XYY individuals in various specific sub-populations. Mental hospitals, mental penal institution and prisons were selected for screening, and not surprisingly, the studies found that an extra Y chromosome predisposes the individual to aggressive and antisocial behavior and is associated with mental retardation. [7]

Signs & Symptoms

Signs

Characteristics of XYY syndrome are often subtle and do not necessarily suggest a serious chromosomal disorder. Thus, males with this condition are often undiagnosed or misdiagnosed. The most common

physical difference is increased height, which usually becomes apparent after the age of five or six, and results in an average height of about 6 feet, 3 inches by adulthood. Some individuals with XYY also develop severe cystic acne during adolescence. Fertility and sexual development are normal. Besides the potential for increased height, most affected individuals typically have a normal physical appearance (phenotype). [8]

Boys with XYY syndrome typically have normal intelligence, although, on average, IQ is 10 to 15 points lower than siblings. Affected boys may exhibit mild delays in reaching developmental milestones. Learning disabilities have been reported in up to 50 percent of cases, most commonly speech delays and language problems. Reading difficulties are common due to an increased incidence of dyslexia. [9]

In some cases, affected individuals develop behavioral problems such as an explosive temper, hyperactivity, impulsivity, defiant actions, or, in some cases, antisocial behavior. There is a higher rate of attention deficit and hyperactivity disorder and a smaller increased risk for having an autism spectrum disorder. [10]

Symptoms

- 47, XYY syndrome typically causes
- Often no unusual physical features.
- Most males with this chromosomal change and are able to father children.
- Taller than average height more than Normal Testosterone levels
- Learning problems

Causes

XYY syndrome is a rare chromosomal disorder caused by the presence of an extra Y chromosome. Normally, males have 46 chromosomes including one X and one Y chromosome. Males with XYY syndrome have 47 chromosomes, two of which are Y chromosomes. Most cases of XYY syndrome are due to a cell division error in the sperm prior to conception. Rarely, the cell division error occurs after conception resulting in a mosaic of cells with 46 chromosomes and 47 chromosomes. The exact cause for why these errors in cell division occur is not understood. [11]

Other Causes of Jacobs Syndrome-

- Serum sickness
- Sweet's disease
- Jacobs arthropathy-camptodactyly syndrome
- Hypertrophic pulmonary osteo-arthropathy
- Rhizomelic pseudopolyarthrits

How does an extra Y chromosome affect a person?

The effect of having an extra Y chromosome can be very varied, but the majority of people with XYY syndrome lead normal lives. They go to ordinary schools, have successful careers, marry, have children and live until old age. Most people with an extra Y chromosome will never know they have this, as they never have a reason to get their chromosomes checked. However, some people with XYY syndrome may experience problems such as learning difficulties and/or behavioral problems. [12]

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 fail to separate when sperm cells are formed. During this process a cell must first replicate its chromosomes so that it has two copies of each and then divide twice to produce four sperm cells. In some cases both of the Y chromosomes will go to the same cell and when this fertilizes an egg (when a baby is made) will produce an embryo with two Ys. It is also possible that a similar event could occur in the very early stages of the development of an embryo. [13]

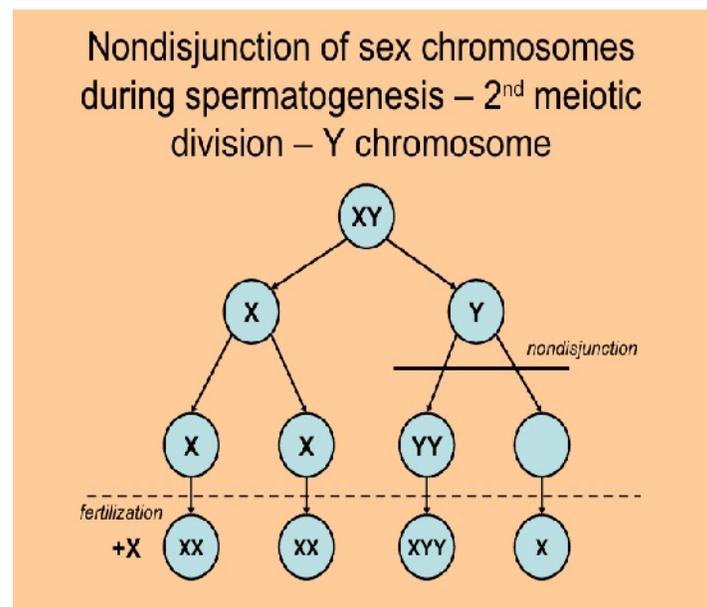


Fig-3: Non-disjunction of sex chromosomes

Complications list for Jacobs's syndrome:

The list of complications that have been mentioned in various sources for Jacobs syndrome includes:

- Acne
- Tall stature
- Aggression

Affected Populations

XYY syndrome is a rare chromosomal disorder



present at birth that affects only males. It is estimated to occur in approximately one in 1,000 live births.

Growing up with XYY

Boys with XYY syndrome grow slightly faster than average in childhood, and their average height is 188cm (6 feet 3 inches). Puberty is normal and at the expected time. Hormones are normal and no special treatment is needed. Homosexuality is no more common than in the general population. Boys with XYY syndrome may go on to further education, but are less likely to do so than their brothers and sisters. Boys with XYY syndrome are not more likely to get a criminal conviction than other boys of the same intelligence level. Boys with XYY syndrome are not more likely to go to prison or young offenders' institutions than other boys. [14]

First year of life

Boys born with XYY syndrome look and behave just like other babies. They are not unusual in weight or length. Problems at birth are no more common than in any other child.

Age 1 to 5 years

Boys with XYY syndrome usually sit, crawl and walk at the usual time, but they may be slower to learn to talk than the average child. About half of boys with XYY syndrome have some delay in speech. In these boys, speech therapy can be helpful, starting by around 3 to 4 years of age. With help, the problem usually disappears after a few years. [15]

Boys at school (5 to 16 years of age)

Boys with XYY syndrome tend to be taller than other boys of their age. This can mean that more is expected from them than from other children of the

same age. They have a wide range of abilities, just as all children do. The majority of boys with XYY syndrome are within the normal range for intelligence. However, research has shown that the average intelligence of boys with XYY syndrome is slightly lower than that of boys with the normal number of chromosomes. It is possible to have above average intelligence and have XYY syndrome. About half of all boys with XYY syndrome may need some extra help at school, but the majorities manage well at mainstream school. When there are learning difficulties, these tend to involve language and adapting to the school environment. [16]

Some may have behaviour problems such as temper tantrums, which may require specialist support. These problems usually respond well to treatment. Some boys are described as easily distractible and more active physically. If parents are aware of this and can channel the child's energies constructively, this does not need to be a problem. Aggression is not more frequent than in other children. The boys seem to cope less well with conflict and stress than their unaffected brothers and sisters. [17]

A strong, supportive home environment is especially important for affected boys. There is no increased risk of any serious mental illness.

Adulthood

The majority of men with XYY syndrome, around 75%, are in employment, in a wide range of jobs. Just as many men with XYY syndrome marry, have normal sex lives and have children, as men with the normal number of chromosomes. They also do not appear to be at any higher risk of having a son with XYY



syndrome themselves. [18]

Key features

- Normal appearance, typically tall stature
- Intelligence usually in the normal range, but an increased need for educational support especially with reading and writing
- Increased vulnerability to ADHD (attention deficit hyperactivity disorder)
- Increased vulnerability to autistic spectrum disorders
- Perhaps increased risk of asthma and epilepsy

Main physical features

Height: tall

Boys and men with XYY are often but not always taller than their peers. It is common for them to reach 6'3" (1.88m) and taller. There is some uncertainty about when the extra height becomes apparent, but two recent studies including a total of 134 boys and men both found that height was normal until approximately 6 years of age and then began to increase; at 11 years almost all boys were taller than average and at 13 nearly all the boys were significantly taller than their peers. Most boys over 6 years were in the tallest 15% of boys for their age and most over 13 were in the tallest 2%. All of the boys had normal proportions and most were a normal weight for their height, although some boys in a recent American study showed a tendency towards overweight around the midriff (central adiposity) (Ratcliffe 1990; Geerts 2003; Ottesen 2010; Bardsley 2013).[19]

A survey of unique families gave a slightly different

picture. 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 noticed 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 (Unique).

Teeth

Boys with XYY are likely to have larger teeth with longer roots than other boys. They can also have a forward-jutting lower jaw and under bite. Dental problems have been found in approximately 1 in 5 boys (20%). Unique parents reported some kind of dental problem in a much larger number: 3 boys out of 4 (14/19). The problems include failure of first (milk) teeth to fall out; overcrowded teeth; poor enamel quality; and missing teeth. Some of these dental concerns may not be explained by the extra Y chromosome. Some boys had teeth removed due to overcrowding; others wore braces to correct the dental position.

Genitals

A large recent review found that most boys regardless of age will have enlarged testes but researchers found no evidence of increased hormone levels linked to this, and are unsure what this finding means. The same review found no increase in minor genital anomalies such as undescended testicles or hypospadias, where the hole usually at the end of



the penis is on the underside instead (Ross 2009; Bardsley 2013). A review of Unique members aged 5 – 18 suggested that enlarged testes may be less common than the research suggests: just 3/21 families confirmed this feature, although the increase in size would not be something most families are aware of.

Medical issues

Asthma

Most boys with XYY do not have asthma, but it is more common than in the general population. Asthma was found in almost 39% (35/89) of boys in one large recent study, compared to a general population level of 9.6% (Bardsley 2013). The authors point out that the extra Y chromosome may be amplifying the risk of atopy, as there is a greater prevalence of atopy/asthma and a reduced relative airway size in boys compared with girls. The Unique 2014 survey showed asthma at a lower frequency in 5/20 boys, of varying severity, but all boys were using inhalers, and one had been repeatedly hospitalized with asthma. Unique also found a high rate of chest infections in the first three years of life or later in childhood, with 14/21 families reporting this.

Seizures

There is a possibility that seizures are more common in boys with XYY than in the general population. The most comprehensive study to look at this found that among boys diagnosed prenatally, 1/35 had seizures, giving a rate of 3% compared with 1% of unaffected boys. They are treated with standard anti epilepsy drugs. The Unique 2003 survey also suggested that among this highly selected group, seizures were 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, and can be explained at least in part by the fact that boys with epilepsy are more likely to have their chromosomes examined. Two boys outgrew them in adolescence and there were no reports of seizures occurring in adults.

Do men with XYY die younger

Two large follow up studies of men with XYY have revealed that they are likely to die younger than other men. One study of all men diagnosed with XYY in a single country, Denmark, showed that life expectancy for men with XYY was 10 years 9 less than for others, with men with XYY dying on average at 68 years rather than 78. Both studies showed an increase in deaths from respiratory diseases, and the Danish study also showed more deaths from cancer, neurological diseases and accidents.

- Fine motor control
- Low muscle tone

A number of studies have looked at motor control and coordination in boys with XYY. Long term screening studies found that in general boys had reduced fine motor coordination, meaning that boys were more likely to face problems with tasks such as writing, drawing and cutting with scissors, as well as balance. In two later studies on prenatally diagnosed groups parents reported motor delay or lack of coordination in 25-35% (3/12 and 4/15) of boys. Later studies assessed this independently and found that the XYY boys performed slightly less well on tests of strength,



speed and dexterity (Ratcliffe 1999; Ross 2009; Leggett 2010). Mild tremors were common and could be seen while resting or during movement (intention tremor) in 43% (39/90) boys in the largest cohort study. In most boys the tremor is too mild to affect writing. Low muscle strength and tone was also noted in half of the 35 boys prenatally diagnosed with XYY in this study. While these symptoms are usually mild they might impair handwriting or other skills and if this is the case extra support in school or occupational therapy might be needed. Other studies confirm these findings, showing intention tremor as both more frequent and more intense in XYY. These observations are amply supported by a Unique survey from 2003, in which 36 families (84 per cent) recorded a problem with either fine motor control or balance (Unique). Unique survey in 2014 again confirmed these findings, with boys diagnosed during childhood more likely to be affected. The low muscle tone had multiple effects, on balance, posture, stamina, sports activities, as well as on writing. Tremor was less common than in the studies, with only 2/21 boys having a hand tremor.

Educational features

Speech and Language

Speech and language is a common area for concern, and having an extra Y chromosome increases the likelihood that a boy will have speech and language difficulties. Small early studies suggest speech delay in up to half of all boys with XYY, making it more than twice as common as in boys without an extra Y chromosome (Ratcliffe 1999; Geerts 2003). Larger, more recent studies show that some speech

problems were found at all ages - although not in all boys - and that at school boys tended to struggle with language and verbal tests, even though IQ (intelligence quotient) was typically in the normal range. In particular they struggled with figurative or ambiguous language, expressing them. [20]

Strengths and weaknesses

Learning strengths often mentioned by parents included mathematics, science, art, visual learning, practical subjects (woodwork, cookery) and, when interested, curiosity and memory. Slightly less common strengths include physical activities (but not necessarily team or contact sports), and spatial awareness. Weaknesses include, most often, literacy, including both reading and writing, motivation (very hard to persuade to do something), coping with pressure, and above all, concentration and staying on task.

Almost every boy had some difficulties with focus and attention, and needed support to keep up his concentration. Learning tasks were best when short, with frequent breaks. Specific writing difficulties were also common.

Behaviour issues

It is generally agreed that boys with XYY are vulnerable to easy distractibility and hyperactivity. Multiple studies of both boys diagnosed before birth and after have shown raised rates of attention deficit hyperactivity disorder (ADHD), and these are supported by parental reports. However, when does energetic become overactive? And when does overactive become hyperactive? In the group of 57 boys tested in the UK and eventually reported in the



medical literature by Bishop 2011, hyperactivity and inattention were indeed common, found in around one third of boys regardless of when they were diagnosed. When considered in more detail prenatally diagnosed boys tend to be described as restless, hyperactive or inattentive, and were often described as busy and energetic. Overall they were described as energetic boys who may have difficulty concentrating. [21]

Behavior difficulties

Sexual behavior

Unique receives occasional queries about sexual behaviour, particularly in adolescents with XYY. We have no information to suggest that sexual behaviour is any different to adolescents with XY.

14 years

Early reports of criminal convictions. The great majority of men with XYY lead law abiding lives, do not behave antisocially and do not have criminal convictions. Early reports of men in penal institutions led to a belief that raised testosterone levels in men with XYY leads to aggressive behaviour. This research should be viewed with extreme caution, because it relies on small, selected groups, and other studies found normal testosterone levels in boys and men with XYY. Follow-up studies of men with XYY have indeed found an increase in some criminal convictions. [22]

Puberty

Repeated studies have shown that puberty is generally little different in boys with XYY to boys with XY. One of the early studies found that it started on average 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. In a larger and more recent study, puberty in some boys occurred early in the age range of childhood

XYY: The Criminal Gene

Additionally, this data suggests that men with an extra sex chromosome may be more likely to commit sexual crimes than other criminal offenders. The clinical features common to both groups are tall stature and low or "low normal" intelligence. Because tallness does not seem to predispose individuals to criminal behavior, it appears the intelligence defect and accompanying abnormal features of the central nervous system are the most likely causes of the XYY's tendency to commit crimes.

Their personalities show extreme instability and irresponsibility, and in their criminal behaviour these men do not appear to have considered any but the most immediate consequences of their actions. They have few constructive aims for the future and the plans they make are generally unrealistic. In their emotional responses they show very little depth of affection for other and their capacity for understanding is more limited than would be expected from their level of intelligence. They display an impaired awareness of their environment, which appears, at least partly, to account for their inability to respond.

1. Organic disease: hormone imbalance

Early research indicated that traits often associated with excessive hormone secretion, such as aggressive behavior, tallness, subnormal intelligence



and acne, may also be characteristics of the XYY individual, because the Y chromosome is the male sex determinant, the presence of two such chromosomes may result in a "supermale" with above normal hormone levels.

According to one researcher, excess amounts of plasma testosterone, the hormone principally responsible for the development of the secondary male sex characteristics, exists in certain XYY individuals. If it can be shown that an extra Y chromosome causes this hormone to exist in abnormal amounts, then the hypersecretion of this hormone, which controls the degree of aggressive behavior, may be the vehicle that translates the XYY abnormality into antisocial behavior. Such information is vital to understand the XYY individual and clarify the link between today's medical and technical advances and the impact such advances have in the legal arena. For example, postpartum disorders are now being recognized as valid defenses in cases of infanticide.

These disorders, which range from mild depression to a complete disassociation from reality, are also believed by many medical experts to be caused by tremendous upheavals in hormone levels after childbirth. The fact that not all XYY individuals develop a criminal history, or that not all new mothers experience postpartum psychosis or depression, does not negate the fact that many individuals are affected by such disorders.

2. Competency to stand trial

An XYY individual who commits a crime may have been unable to control his actions at the time of the

crime, but the individual may not be incapable of understanding the proceeding against him and assisting in his defense. Even if he were found incapable, the antisocial behavior that may have accounted for the crime is not necessarily of an ongoing, uncontrollable nature. The XYY individual, although possibly hampered by permanent mental deficiencies and learning disabilities, usually regains control of his emotional faculties. Because most jurisdictions require a competency determination within eighteen months of indictment, if the defendant's only mental disease or defect is the temporary loss of emotional control.

3. Recommendation

Rehabilitation focuses on the individual who repeatedly commits crime. This theory is based on the rationale that criminals endanger society, and that their criminal propensities must be eliminated before they can be returned to society. The problem with applying this theory to XYY individuals is that their criminal propensity is caused by a genetic abnormality that cannot be eliminated. The theory may, however, be useful in reducing the criminal propensity of the XYY defendant by providing support programs like protective confinement, home monitoring, mandatory periodic supervisory examinations and required enrollment in schools with controlled environments. It appears, therefore, that if the XYY syndrome is considered in sentencing—especially in light of the rehabilitative theory of punishment—society's interest in supervising the XYY individual and the XYY individual's interest in overcoming the aggressive tendencies created by his



abnormality will both be served. [23]

A Genetic Causes of Criminal Behaviour

The XYY chromosomal deviation, premenstrual syndrome, the male hormone correlation with violent crime, biological origins of schizophrenia and alcoholism, temporal lobe epilepsy, and biochemical triggering of hostile behavior. The discussion then reassesses the legal system's traditional concepts of guilt and punishment in relation to the scientific findings presented. Some issues considered are the development of a genetic or biological defense that parallels the insanity defense to demonstrate the absence of criminal responsibility and the use of new rehabilitative and preventive procedures to counter genetic and physical states contributing to deviant behavior.

The XYY Supermale and the Criminal Justice System

An individual who possessed certain physical characteristics could not be expected to refrain from crime unless the circumstances of his or her life were exceptionally favorable. The belief that criminality may be at least in part, genetically predetermined or influenced by biological characteristics, has been resurrected by a new awareness and sensitivity in the fields of medicine, psychiatry and psychology.

Today the legal profession is advancing defenses to criminal conduct based on biological determinants such as postpartum depression and psychosis, 6 premenstrual syndrome⁷ and genetic determinants such as XYY syndrome.

The American criminal law system is founded on the assumption that all individuals are equally able to

perceive what is "right" and to act freely in accordance with that perception the assumption that the threat of punishment will effectively deter most individuals from committing criminal acts, and that sufficient detention in a penal institution can rehabilitate one who has committed a crime and prepare him or her for reentry into society. This Comment will explore the possibility that individuals exist who cannot be as easily "plugged" into the current system of American jurisprudence as can others. This Comment will primarily focus on males who have an extra Y sex chromosome (XYY individuals)," the possibility of inherent antisocial behavior in such individuals' 2 and the inability of the present criminal law system to dispense equal justice to them. This Comment will discuss the nature of the XYY syndrome and its reported characteristics, 14 possible theories of defense for the XYY individual accused of a crime" and other uses related to an XYY syndrome defense including a recommendation for its use in sentencing. The potential danger of XYY identification used not as a shield to protect criminal defendants, but rather misused as a sword against them. [24]

XYY and the Insanity Defenses

The insanity defense originally was created to recognize that some individuals were not morally blameworthy and therefore were not criminally responsible for their actions because they did not understand the moral significance of their acts. A defendant is entitled to an acquittal if, at the time of the crime, he or she was so impaired by mental illness or retardation as to be "insane" within the



meaning of the law.

This test of responsibility is universally called the "irresistible impulse" test, which provides that a defendant is entitled to acquittal on insanity grounds if his or her commission of the crime was caused by an "insane impulse" that controlled the defendant's will. It is not necessary that the defendant's action be sudden, but only that a mental disease caused the lack of control. If the XYY defendant is unable to cope with stressful situations and has great difficulty resisting aggressiveness in his actions, the defect could be responsible for the XYY's inability to control his behavior.

The control test requirements, however, express in absolute terms that nothing less than a complete inability to control oneself will suffice to free the accused from responsibility.

Thus, unless medical evidence can demonstrate that an XYY individual finds it virtually impossible to control his behavior at the time the crime is committed, it is unlikely that the XYY defense alone would warrant acquittal of the individual under the irresistible impulse test." In jurisdictions that employ the control tests, however, the burden of proof with respect to the issue of insanity will determine the significance of XYY syndrome as a successful defense.

Diagnosis

A diagnosis of XYY syndrome is made based upon a thorough clinical evaluation, a detailed patient history, and specialized tests (i.e., chromosomal analysis) that detect the presence of an extra Y chromosome (47, XYY karyotype).

A diagnosis of XYY syndrome may be made before birth (prenatally) through amniocentesis or chorionic villus sampling (CVS). During amniocentesis, a sample of fluid that surrounds the developing fetus is removed and analyzed, while CVS involves the removal of tissue samples from a portion of the placenta. Chromosomal studies performed on such fluid or tissue samples may reveal the presence of an extra Y chromosome.

Clinical Testing and Work-Up

Speech and language assessment should occur during the first 24 months. Reading assessment should occur by school age to rule out dyslexia. Behavioral assessment should be considered for children who are having difficulty with symptoms such as impulsivity, poor attention, or social skills.

Testing Validity

A better, more valid measure of XYY tendencies toward aggressive behavior can be examined with appropriate psychological testing employing outcomes suitable for valid testing. Validity in scientific testing is important. Validity does not mean accuracy. Validity means that a study or experiment tests what it is meant to test. Therefore, if we seek to find out if XYY men are more aggressive than XY men, aggressive behavioral tendencies—not acts of criminal violence and subsequent incarceration rates—should be the measure.[25]

Thus when appropriate and comprehensive psychological testing is implemented, men with XYY syndrome are shown more likely to be aggressive and exhibit more aggressive behaviors than normal XY men



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