Human Behavior and Genetics
W. Schmid and J. Nielsen, editors

SPATIAL ABILITY OF MEN WITH KARYOTYPE 47,XXY, 47,XYY, OR NORMAL CONTROLS

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SUMMARY

The term spatial ability refers to a number of higher-level cognitive processes. Perhaps due to its acknowledged importance for the solution of various performance IQ-tasks, a number of theories have been proposed to explain the expression of spatial ability. Sociological, biological, and interactionistic theories have purported to account for the etiology of spatial ability. One of these theories - the X-linked recessive genetic theory - was subjected to test in the present work. Groups of men with an extra X-chromosome (Klinefelter's syndrome, karyotype 47,XXY), and with an extra Y-chromosome (karyotype 47,XYY) went through a number of spatial tasks, and their performance was compared to the predictions of the X-linked theory for these groups. Spatial ability in women lacking X-chromosome material was also discussed in relation to the theory. In general, the predictions of the X-linked, recessive theory failed to account for the expression of spatial ability in two out of the three groups studied. As the X-linked theory is furthermore unable to explain the results of a number of recent large-scale studies of the familial transmission of spatial ability, it was concluded that the theory must be refuted in its present form. A number of alternative theories were pointed to, and the newest of these - the psychoneuroendocrinological "Optimal estrogen level" theory - was briefly outlined in order to account for the present findings. Finally, clinical and educational implication of low spatial ability in persons with aberrant karyotypes were discussed.

INTRODUCTION

Spatial ability refers to the capacity of an organism to orient itself to the surroundings, to deal with objects changing place in space, and to comprehend patterning of visual stimuli. Spatial ability refers also to visualization capacity, i.e. to the ability to invert, rotate, twist or manipulate visual stimuli. Spatial ability seems quite resistant to cultural influence or more narrow environmental manipulation[1,2,3,4]. Attempts to improve spatial ability by systematic training seem to have little, if any effect. In most studies in which a training effect was found, the impact was transient and did not transfer to other spatial ability

tasks[5,6]. The relative spatial ability score of a given individual within a population has been found to be stable from childhood onwards. However, a repeated observation in spatial ability research is the appearance of an average sex difference just around puberty[5]. At that time males take the lead, apparently not because their spatial ability improves much after puberty, but rather because the spatial ability of postpubertal girls recedes.

Recent years have witnessed a surge in interest in spatial ability, partly because of controversy about its etiology, but perhaps more importantly because spatial ability seems related to a number of essential higher-level human functionings of considerably educational relevance. Thus, a number of studies have demonstrated that spatial ability is important for achievement in engineering, architecture, physics, geometry, musical composing, playing chess, and even in science in general, areas in which adult male dominance often can be observed[5,7,8,9]. Spatial ability is also of interest to the clinician. On the neurological level, it is generally assumed that the processes subserving spatial ability are localized mainly in the right brain hemisphere[10,11,12]. Accordingly, brain damage in the right hemisphere can be expected to cause greater difficulties with spatial ability tasks than left brain damages and has been confirmed[ex.13,14]. It was further observed that right-sided lesions have a more severe effect on spatial ability in adult men than in women[15,16]. Waber[17,18] found that late maturers had a more pronounced right brain lateralization and scored higher on spatial ability tasks than did early maturers.

A number of theories have been advanced to explain the expression of high spatial ability. One of the more prominent genetic theories was first suggested by O'Connor[19] and later corroborated by Stafford[20,21,22]. According to this theory, spatial ability is sex-linked, and is coded for by a recessive gene on an X-chromosome. The theory has the merit of being elegantly simple, and at the same time allowing for a number of precise predictions that can be rigoriously tested.

The X-linked recessive theory predicts that fewer women than men demonstrate high spatial ability. Bock and Kolakowski[23] have suggested that the sex ratio for high spatial ability is about 1 to 2, and that 25 per cent of all men show high spatial ability. The X-linked recessive theory also predicts a certain pattern of familial cross-sex transmission of spatial ability. A number of studies[20,23,24,25,26,27] apparently supported the X-linkage theory of spatial ability. Recently, however, Boles[28] was able to demonstrate that the outcome of most, if not all early studies supportive of the theory, could be attributed to chance variations[see also 4]. Furthermore, a number of later studies gave no support to the X-linked recessive account of parent-children transmission of spatial ability[29,30,31,32], and Bock[33] observed that women high in spatial ability do not always have fathers high in ability. In conclusion the most prominent genetic theory for the transmission of spatial ability was not substantiated by the outcome of the majority of family studies. However, a few studies[26,27] provided moderate support for the theory.

Fortunately alternative research strategies for testing the theory are at hand. One such alternative is to study persons with sex chromosome abnormalities. Studies of spatial ability in women with Turner's syndrome exemplify this approach[34,35]. About half of all women with Turner's syndrome lack one X-chromosome completely and accordingly have karyotype 45,X. According to the X-linked recessive theory for spatial ability, women with karyotype 45,X will manifest as high spatial ability as do normal men, because the recessive allele for spatial ability in both cases comes to full expression[36]. Contrary to the prediction, however, women with karyotype 45,X show extremely low spatial ability[37]. Turner's syndrome further includes women who lack only part of the second X-chromosome. The X-linked theory predicts lower spatial ability in this group than in karyotype 45,X women, because at least a number of these women can be expected to need two alleles of the spatial gene for the expression of high spatial ability. They are in fact expected to have spatial ability quite close to that of normal women, according to the X-linkage theory. Neither of these predictions are confirmed,

however, in that women with Turner's syndrome who lack only part of the second X-chromosome obtain a spatial ability score indiscriminable from that of karyotype 45,X Turner women[37]. Turner's syndrome finally includes women who lack the second X-chromosome in only part of their body cells or are mosaics with isochromosome X. The number of such women tested by Nielsen, Nyborg and Dahl[37] was small, but the evidence at hand showed that these women do not score differently from other Turner's women. It seems fair accordingly to conclude that the results of studies on spatial ability in women who lack more or less X-chromosome material provide no support for the X-linked theory.

Persons with sex chromosome aberrations other than those of Turner's women also lend themselves to testing the X-linked theory. Two such groups consist of men with a supernumerous X- or Y-chromosome. According to the X-linked recessive theory, men with an extra X-chromosome (Klinefelter's syndrome and karyotype 47,XXY) can be expected to obtain lower spatial ability score than normal 46,XY males, and in fact to perform exactly like normal females on spatial tasks. On the other hand, men with an extra Y-chromosome (karyotype 47,XYY) can, according to the X-linked recessive theory, be expected to obtain a spatial ability score equal to that of normal men because they need one allele only on their sole X-chromosome. Following the theory it does not matter for spatial ability whether one or two Y-chromosomes are present in the karyotype.

The present study was designed to test the X-linked, recessive theory by observing whether spatial ability is distributed according to the predictions of the theory in groups of men with 47,XYY and 47,XXY, respectively. Groups of men with these types of sex chromosome aberrations were subjected to a number of tests assumed to draw upon spatial ability, including the Rod-and-Frame test and the Embedded-Figures test[6]; also used in 27,37. The results were compared to those of normal female and male controls. While an important goal of the study was to provide new pieces of evidence to the puzzle about the X-linkage of spatial ability, an equally important perspective was to gain a more adequate understanding of the possibly adverse consequences on cognition of aberrant sex chromosome complement in order to improve the conditions for the innocent victims of these "experiments" by nature.

METHODS

Subjects. 27 men with Klinefelter's syndrome previously studied[38,39,40] were selected to participate in the present investigation. Of these, three made no reply to the invitation, two were dead, one was too ill to be tested, and one had changed address without notice. The remaining 20 men with Klinefelter's syndrome were included in the study. Their age ranged between 15-60 years with a mean of 35 years 4 months and SD 14 years 7 months. Nine of these were diagnosed during admission to (1) psychiatric wards, three to (2) forensic-psychiatric wards, three to (3) neurological units, and five to (4) medical departments, and one was found in a population study. 27 other men with karyotype 47,XYY previously studied[38,41,42] were selected. One of these refused to be tested, one had moved abroad, one could not be found at his last address, and the last man had participated in so many previous studies that it was found adviceable not to strain him by further investigation. The remaining 23 men with karyotype 47,XYY were tested. Their age ranged between 16-53 years, with a mean of 27 years 8 months and SD 10 years 6 months. Five of these men were diagnosed during admission to (1) psychiatric wards, eleven to (2) forensic-psychiatric wards, two to (3) medical departments, and the last five came from a population study. Male students and sisters of Turner's women acted as controls.

Instruments and procedure

Spatial ability was measured individually by means of the Rod-and-Frame test, the Embedded-Figures test, the Human-Figures-Drawing test, and by Money's Road-Map test of direction sense.

The Rod-and-Frame test apparatus (RFT:[43]) used was a transportable model from DARRO Scientific. It consisted of a table-top sized box. The subject put his head into one end of the box so that his view was restricted to the inside of the box. A square frame with a movable rod inside it was visible at the other end of the box. The frame was tilted 28 degrees to the right or to the left of gravitational vertical. The subject's task was to adjust the rod to apparent vertical within the stationary tilted

frame. The traditional method of scoring the RFT[43] gives an unsigned, unweighted deviation score (USD). It disregards information about the direction of the deviation and gives no estimate of the response consistency of the subject. Therefore, in addition to the traditional method a new method of scoring the RFT was used as described in detail elsewhere[44,45,46]. The direction of deviation of the rod from gravitational vertical was recorded to calculate a "signed deviation" score. An account was kept of whether the rod was adjusted to the same side to which the frame tilted or to the other side. The "pure" effect of tilt of the frame on the final position of the rod (the frame dependence parameter, φ) was calculated from the data on signed deviation of rod setting, in degrees, from gravitational vertical.

The Embedded-Figures test (EFT:[6]) was the short form of 12 figures by Jackson[47] administered the traditional way. The subject was required to find the twelve simple geometrical figures "embedded" in more complex figures. The score was the mean number of seconds per figure required to find the simple figures.

The Human-Figures-Drawing test (HFDT) was scored according to Witkin et al.'s[6] "Articulation-of-body-concept" scale said to reflect the extent to which the drawings mirror the body. The adult version of the scale (the so-called ABC scale) was used. The degree to which the drawing had definite limits (boundaries) and the extent to which the "parts" within limits were separate although kept together in a "gestalt" was judged in a single global evaluation based on a number of specific criteria that were read directly from observable characteristics in the drawings rather than on the basis of a more traditional projective interpretation. After all identification marks had been removed, the drawings were rated by two independent judges with previous experience in evaluating drawings. They looked for three main indicators of degree of articulation: (1) fair integration of reasonably formed parts of the body and articulation in representation of body and face expressions; (2) identity of sex differentiation where the role and the articulation of the sex characteristics of the figures drawn were evaluated, and finally (3) the degree of details in the drawings. Scores ranged from 5 for the most primitive, immature, undetailed and disintegrated draw-

ings to 1 for the most articulated drawings with emphasis on many details in face, head, clothing, expression, forms and sex characteristics consistent with rational integration of body and clothing. In case of disagreement in the estimation of the drawings, the two judges solved the problem by talking to agreement.

Money's Road-Map test of direction sense (MRMT) was used to study direction discrimination. It requires orientation to right and left simultaneously with orientation towards and away from the subject. The test consists of a flat surface resembling part of a city map. Heavy lines on the map indicates a standard test route which is laid out in such a way that there are eight different turn types (turning right after going up, down, right, and left; turning left after going up, down, right, and left) with a total of thirty-two turns. A shorter, heavy line indicates a preliminary practice route of three turns. Both practice and test routes were traced on the map by the examiner while the subject looked on. The subject was then requested to retrace the routes saying whether he turned right or left at the corners[48,49]. The subjects were corrected for errors on the practice route only. The score was the total number of left-right errors made on the test route.

RESULTS

As shown in Table 1, men with karyotype 47,XYY obtained the lowest spatial score of the groups tested, followed by men with karyotype 47,XXY. Normal female controls had higher spatial ability than these two groups, but lower spatial ability than normal male controls. Analysis of variance (ANOVA) showed that for four tests the differences between the four groups were significant at at least 5 per cent level. Money's task failed to demonstrate significance. The patterning of the data is consistent. Whether the Rod-and-Frame test was scored according to the traditional method or by the new method, men with a surplus of sex chromosome material demonstrated a much lower spatial ability (field dependence) than did normal men while the level of spatial ability for women lay in between. This trend could also be observed in the two other indicators of so-called field dependence, i.e., the Embedded-Figures test and the Human-Figures-Drawing test.

TABLE 1

SPATIAL ABILITY OF MEN WITH KARYOTYPE 47,XYY OR 47,XXY OR NORMALS

Groups	n	Rod-and-Frame test score ad modum Witkin (degrees)		Rod-and-Frame test score ad modum Nyborg (φ) (degrees)		Embedded-Figures test score (seconds per figure)		Human Figures Drawing test ad modum Witkin (ABC-scale)		Money's Road-Map test of direction sense (Number of errors)	
		Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD
Men with karyotype 47,XYY	23	7.84	7.36	6.81	7.03	103.92	49.59	3.44	1.27	4.91	5.57
Men with karyotype 47,XXY	20	5.92	4.49	5.12	4.89	91.55[c]	39.73	3.00	1.45	2.00	4.38
Normal women with karyotype 46,XX[a]	19	4.59	4.70	3.97	5.09	61.25	23.68	2.53	.84	1.68	4.57
Normal men with karyotype 46,XY[b]	14	2.33	1.74	1.33	2.17	47.90	30.16	1.83	1.02	3.14	2.80
Significance		$F(3,72)=3.41$; $p = .02$		$F(3,72)=3.18$; $p = .03$		$F(3,72)=8.36$; $p = 0.0001$		$F(3,72)=5.85$; $p = .0012$		$F(3,72)=2.16$; $p = .099$	

[a] Sisters of women with Turner's syndrome (after[37])
[b] Nyborg (unpublished data)
[c] Significantly different from 46,XX; $t(31.25)=2.91$; $p = .007$ (fractioned D.f. due to inhomogeneous variance prompting separate variance estimate).

Scores in Money's Road-Map test for direction sense also followed this pattern although the unusual high score in normal 46,XY's cannot be accounted for. T-tests indicated that men with karyotype 47,XYY scored significantly lover on all spatial ability tasks except Money's Road-Map test than did normal men. Men with Klinefelter's syndrome consistently obtained lower spatial ability scores than did normal female controls in the tests applied, but the difference was significant only in the Embedded-Figures test ($t(31,25)=2.91$; $p<.007$). Due to inhomogeneous variance, a separate two-tailed variance estimate was made, as reflected in the fractionated D.f.'s in Table 1.

DISCUSSION

Numerous reports on general intellectual capabilities in men with karyotype 47,XYY have appeared since Sandberg et al.[50] first documented the existence of men with an extra Y-chromosome. Normal or even superior IQ is not incompatible with karyotype 47,XYY[51,52,53], but a tendency for a slightly depressed IQ score has been observed in most men with an extra Y-chromosome[41,54,55,-56,57,58,59,60]. It is to be noted, however, that men with an extra Y-chromosome are sometimes recruited from psychiatric institutions or forensic-psychiatric wards for study. As such populations might possibly obtain lower IQ scores than do unselected groups, general conclusions about a reduced mental capacity in men with karyotype 47,XYY based on such material might be unjustified. However, Nielsen and Christensen[41] compared the mean IQ of 17 institutionalized men with karyotype 47,XYY to 9 non-institutionalized men, and found no difference, while Witkin et al.[61] screened a Danish population of army conscripts for XYY men, and observed a significant IQ depression in this group. Thus, a slightly depressed general IQ has been found in men with karyotype 47,XYY, whether these men were drawn from institutions or not.

Performance IQ correlates positively with spatial ability scores. It is therefore of interest to note that while verbal ability seems depressed in men with an extra Y-chromosome, their performance IQ also tends to be slightly below normal. By analogy, we expected spatial ability to be only slightly below normal in the present population of XYY men. However, men with karyotype

47,XYY performed exceptionally poor on all spatial tasks. In fact, only women with Turner's syndrome have ever obtained a lower group mean score on a battery of spatial tasks[37] than have our XYY group. Since the present study is overrepresented by men diagnosed in forensic-psychiatric wards, population bias perhaps could explain the observed low spatial ability. However, a comparison of the eleven previously institutionalized males to the five unselected XYY men revealed no differences: both groups scores equally low. In an independent study, Theilgaard also observed that non-institutionalized men with karyotype 47,XYY obtained a much lower spatial ability score than did matched controls. Thus the study by Theilgaard and the present study both indicate severe inhibition of the expression of spatial ability in men with an extra Y-chromosome.

According to the X-linked, recessive theory, all groups of men with only one X-chromosome should on an average be equally high in spatial ability, in that the theory considers the number of Y-chromosomes present in the karyotype not to influence the expression of spatial ability. We found, however, a pronounced spatial deficit in the group of men with double Y relative to normal controls. Our observations further questions one of the basic assumptions of the X-linkage theory, namely, that gene(s) on the X-chromosome alone conditions the expression of spatial ability. Bock and Kolakowski[23] suggested that in addition to the X-linked component, spatial ability might also be influenced by an autosomal gene. However, this additional hypothesis does not save the theory, because it still cannot explain the present observation of extremely low spatial ability in men with an extra Y-chromosome compared to normal male controls.

A number of studies indicate that the mean full-scale IQ in males with Klinefelter's syndrome is significantly below that of males with normal karyotype[38,54,58,59,61,63]. Significant differences between verbal and performance IQ is not usually found, but there is a tendency for Klinefelter's men to obtain a slightly higher performance than verbal IQ score. In addition,

Cohen's[64] Freedom from Distractibility factor score was low in some of the studies on Klinefelter's men[e.g.39,65]. It seems unlikely that the depression of IQ in men with Klinefelter's syndrome can be explained alone on the basis of a non-random higher proportion of institutionalized men studied, because Nielsen et al.[39] and Witkin et al.[61] both found a substantially lower mean intellectual index in non-institutionalized males with 47,XXY than in hypogonadal males with 46,XY and normal controls, respectively. Acknowledging the correlation between low performance IQ and low spatial ability, we expected men with Klinefelter's syndrome to obtain a spatial ability score slightly below that of normal men. All five spatial ability measures in Klinefelter's men were, however, lower than those of normal females. In one task (Embedded-Figures test) the difference reached significance (see Table 1). Our result thus provides little support for the X-linked recessive theory for spatial ability.

Judged by all available evidence, the X-linked theory is presently in a rather weak position. Positive evidence for the theory is scanty: 1) some early family studies conformed to the predictions of the theory, 2) a later linkage-study indicated modest support, and 3) confirmatory sibling-correlation were found in one study. Negative evidence seems on the other hand to accumulate: 1) it has been demonstrated that the results of the early studies could be ascribed to random variation, and later large scale studies revealed no signs of X-linkage for spatial ability, 2) daughters high in spatial ability have not always eminent fathers, 3) women with Turner's syndrome and men with karyotype 47,XYY have abnormal low spatial ability, and 4) men with karyotype 47,XXY have slightly lower spatial ability than have normal women. It seems therefore fair to conclude that the X-linked recessive theory for spatial ability most probably has to be refuted in its present form.

A number of socialization theories have been proposed to explain development of spatial ability[67,68 and others]. There is, however, a number of problems with these theories. Most, if not all socialization theories assume that the way children are reared or trained affects spatial ability. The way boys are typically reared is said to further spatial ability development while the

rearing of girls hampers their development. It has been shown, however, that boys and girls are not treated as differently as is usually believed[5]. Also the mechanism mediating the postulated socialization effects on spatial ability are left unexplained. Next, socialization theories cannot explain why the sex difference in spatial ability does not show up before puberty, or why the difference becomes so large so quickly at puberty, or why the difference stabilizes shortly after puberty.

Most socialization studies have based their conclusions on correlations in which it is difficult to know what is cause and effect. A perhaps more serious problem with socialization theories is that the notion of differential socialization of boys and girls begs the question of *why* the differential treatment of boys and girls came into existence in the first place. Could it not as well be that boys and girls invite to differential treatment, and that this invitation is willingly responded to by the parents[69]? In that case, boys and girls are treated differentially, not primarily because of prevailing social prescriptions, but rather because parents are reinforcing an already existing disposition in the children. Such a predisposition for differential treatment in boys and girls might be genetically conditioned and was perhaps created by evolutionary forces. Such speculation should of course be replaced by evidence, but their likelyhood challenges socialization theories of spatial ability.

Other theories have been advanced to explain the development of spatial abilities. Some theories consider the role of sex hormones. Already in 1968 the Broverman group[70] noted certain relations between plasma sex hormone concentration and automatization behaviour which probably contains spatial components. They formulated a theory of central nervous activation/inhibition as a function of the estrogen/testosterone balance. The theory ran into a number of difficulties[71,72], and seems not to be generally accepted today. The observations of the Broverman group on the hormone-spatial ability relationship remain, however, and deserve an explanation. There is, in fact, a steadily growing body of evidence pointing to the importance of considering hormones as instrumental for the expression of spatial ability in humans[5,73,74,75]. In addition, animal studies have produced results that

bear a striking resemblance to some of the observations on humans. Thus, male rats usually are superior to females in finding their way in complex mazes. However, neonatal androgen treatment improves spatial performance in female rats, while neonatal estrogen treatment makes spatial ability in male rats deteriorate[76,77]. Similarly, if adult male rats are treated with estrogen their maze learning will be inhibited. At puberty the number of errors made in maze learning tasks increases in female rats. Although only working hypotheses can be generated from animal studies with regard to humans, it is nevertheless thought-provoking to observe that postpubertal human females make greater errors in spatial ability tasks than they did shortly before puberty[e.g. 6, note 1]. As spatial ability components contribute to ability in mathematics and geometry[78,79; see also 5,80], a similar postpubertal decrease in mathematics and geometry achievement can be expected, and has as previously mentioned been observed, but only in females. Recently, Nyborg[note 1] reviewed the literature on a possible relation between spatial ability and sex hormones in humans. The review prompted an attempt to interprete the observations within the framework of a hormonal model. Briefly, it was proposed that spatial ability is coded for by genes on autosomal chromosomes, the expression of which are related to plasma hormone concentrations. Accordingly, the theory assumed that spatial ability is sex-limited rather than sex-linked. As estrogen has been found to have major impact on spatial ability in animals as well as in humans, this biologically very active hormone was considered to be a main factor, while testosterone was said to modulate the effect of estrogen. It was further assumed that "too high" or "too low" plasma estrogen values are equally damaging for the expression of spatial ability, while an intermediate estrogen level maximises the trait. The "Optimal-estrogen-level" (OEL) theory finally assumed that most adult men are slightly below the optimal level, while most adult women are above. More details on the theory can be found in Nyborg[note 1]. Suffice it here to present a few illustrations of how the theory accounts for a number of observations. The absence of a sex difference before puberty in spatial ability can be accounted for by the absence of notable differences in plasma values during

that period. The inhibition of spatial ability in pubertal women can be explained by the considerable surge in their plasma estrogen. The theory also explains the observation that spatial ability is lower in women in the estrogen-high phase of the menstrual cycle than in the low estrogen phase[81], and perhaps provides an explanation for the finding that distinctly feminine women have lower spatial ability than have androgynous women[67,82,83]. The OEL theory can also account for the observation that short-term cyclic estrogen therapy apparently restores the abnormal low spatial ability in women with Turner's syndrome to a level not discriminable from that of their matched sisters, while long-term estrogen treatment was associated with spatial ability as low as in untreated Turner's women[62]. According to the OEL theory, the average adult man has a slightly below optimal level of plasma estrogen for the expression of spatial ability. Increasing the estrogen level slightly leads to full expression of spatial ability as in androgynous men[see 5,73], while an unusual surge in estrogen leads to inhibition of spatial ability as in feminized men with Kwashiorkor syndrome[84] and in feminized 46,XY men insensitive to their own testosterone[23,85]. Accepting the hypothesis that testosterone acts centrally only if conjugated to estrogen (the conversion hypothesis: [86,87]) it becomes understandable that unusual high levels of testosterone also can suppress cognitive ability. In this way, the OEL theory provides an explanation for why the Broverman group[88,89] observed that serial subtraction was optimal at a moderate level of plasma testosterone administered by veneal infusion, while higher levels depressed performance. Granted that testosterone can masculinize the body by peripheral effects and can be converted centrally to estrogen, the OEL theory can also explain the observation that spatial ability is lower in distinctly masculine looking men (i.e. heavy muscles, coarse beard, broad shoulders, little fat tissue etc.) than in androgynous looking men[90,91,92].

The results of the present study can be explained by the OEL theory in the following way. Abnormally low concentration of plasma testosterone (or perhaps abnormal hormonal metabolism) results in physical feminization of men with Klinefelter's syndrome, transgression of the level of estrogen optimal for the

full expression of spatial ability, and suppression of the autosomal gene for this trait. An analogue is perhaps seen in genotypically normal men with feminized bodies due to androgen insensitivity or to Kwashiorkor syndrome. Application of the OEL theory to the present findings in men with karyotype 47,XYY is less clear. This is so, because some studies found men with double Y to have higher than normal levels of testosterone[e.g.93,-94,95], while others found normal testosterone levels[e.g.96,97] and still others found low levels[e.g.98,99]. However, one study noted abnormal low estrogen values in men with 47,XYY[100]. The considerable scatter in sex hormone concentrations observed in men with 47,XYY makes it unclear whether spatial ability is depressed by abnormal low estrogen values and/or by unusual metabolism of testosterone. In both cases spatial ability would according to the OEL theory be depressed.

Clearly, this account of our observations on spatial ability in men with karyotype 47,XXY and 47,XYY needs experimental testing before definite conclusions can be made. Most probably, the OEL theory is much too simple, since hormonal systems interact in many intricate ways. The nature of hormone metabolization, differential sensitivity of the target-organs, prenatal presetting of later sensitivity and many more factors may play an important role. Only further studies will show more details about relations between hormones and spatial ability.

It is generally found that men with karyotype 47,XXY and with 47,XYY achieved less well in school than the average child and this could be due to general behavioral problems as well as to specific cognitive problems. Boys with Klinefelter's syndrome often lack energy and fail to concentrate, while boys with karyotype 47,XYY demonstrate a tendency for hyperactivity as well as failing concentration. Therefore, early diagnosis of these karyotypes seems called for, in order to be better able to provide adequate advice to parents as well as to the children themselves, and to actively intervene with school officials and others in order to promote support if considered necessary. Boys with aberrant sex chromosomes as well as girls with Turner's syndrome and with triple X-syndrome may benefit from informing their teachers that some of them might have special difficulties in

school subjects calling for spatial abilities. These individuals can then receive improved instruction, perhaps by applying learning materials that emphasize serial learning rather than holistic, global learning as is required by diagrams and by the "new math". Perhaps hormone treatment will also benefit some individuals with aberrant sex chromosomes. It is of interest in this regard to note that Annell, Gustavson and Tenstam[101] found a beneficial effect of testosterone treatment on learning in 10-11 year old boys with Klinefelter's syndrome, and that a group of 15-16 year old Klinefelter boys presently in testosterone treatment in our laboratory do as well in school as other boys[66]. Thus, there may be good reason for optimistic prospects for future academic achievement of individuals with these abnormal sex chromosome complements, provided that they receive proper social and educational stimulation in addition to medical treatment.

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