

CYSTINURIA: AN INVESTIGATION OF  
SUGGESTED PSYCHOLOGICAL COMPLICATIONS

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Abstract

The relationship between Cystinuria, a genetic disease, and various psychological variables was examined. It was predicted, on the basis of their over-representation in mental institutions, that Cystinurics would be less intelligent and would exhibit poorer general life adjustment. It was also predicted that Cystinurics would be found to be more external on the internal-external continuum. Subjects included 26 Cystinurics, 12 of whom were compared with matched controls (non-cystinuric siblings). Virtually all of the predictions were rejected. It was concluded that non-institutionalized Cystinurics do not have a greater tendency to suffer psychological difficulty. Two possible explanations of the present findings were then discussed. A follow-up study on families where Cystinuria has already been associated with mental disorder was suggested.

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## Introduction

Cystinuria is a hereditary disorder associated with the excretion of large amounts of the amino acids cystine, lysine, arginine, and ornithine, in the urine (Harris, 1959). Since cystine is poorly soluble in water, its increased concentration in the urine often leads, through precipitation, to the formation of renal stones which are composed almost entirely of cystine. Until recently, the disease, apart from its propensity to cause renal stones, was considered benign. However, recent reports (e.g., Efron, 1965) have suggested that Cystinurics are over-represented in mental institutions, thus raising the possibility of previously unsuspected psychological complications. It was felt, therefore, that a systematic psychological investigation would be illuminating.

### Physiology and Heredity of Cystinuria

Urinary cystine stones were first identified by Wollaston, in 1810, who also recognized the disorder to be familial and somehow genetically determined. Garrod (1908), in reviewing the early literature, concluded that Cystinuria was due to a genetically determined block in cystine metabolism which could also be associated with impaired metabolism of lysine and arginine. Dent & Rose (1951) suggested on the basis of accumulating evidence, that the four amino acids were reabsorbed from the kidney glomerular filtrate by a common mechanism of active transport across the renal tubules. They further suggested that this mechanism might be defective in Cystinuria. Until recently (e.g., Harris, 1959), it was generally believed that Cystinuria could, in fact, be

attributed to this specific defect in renal tubular reabsorption. There was no evidence that any metabolic abnormality existed. Milne, Asatoor, Edwards & Loughbridge (1961), followed by Thier, Fox & Segal (1964), London & Foley (1965), and others, demonstrated that a defect of intestinal absorption of basic amino acids, including Cystine, occurred in association with Cystinuria. More recently, Scriver & Whelan (1971), citing Blasberg & Lajtha's finding (1965) that the amino acid transport systems of brain and kidney are similar, suggested a possible cerebral defect of absorption.

Although it has been established that Cystinuria is associated with defects of amino acid absorption at two sites, kidney and small intestine, the precise nature of the underlying physiological and biochemical processes remains to be fully explained.

The hereditary aspects of Cystinuria have received considerable attention. Harris (1959) concluded that on the basis of previous studies (Harris & Warren, 1953; Harris, Mittwoch, Robson, & Warren, 1955), there exist two distinct genotypes in the disease, a completely recessive (Type 1) and an incompletely recessive (Type 2) genotype. In virtually all cases the parents of Cystinurics are described as heterozygous for the disorder. This means that in the chromosome pair involved in Cystinuria, each chromosome with only one gene-site at which the Cystinuria variant can occur, one of the sites is normal while the other carries one of two possible abnormal genes for Cystinuria, Type 1 or Type 2. The Cystinuric is homozygous for the disorder, that is, each of the pair of chromosomes carries the same abnormal gene, (Type 1 or 2). The parents of Cystinurics with the completely recessive trait have normal urine. The parents of individuals with the incompletely recessive trait excrete increased amounts of cystine and lysine, though less than in the

homozygote. In other words, the Type 1 variant remains unexpressed as long as one of the two genes remains normal, while the Type 2 variant achieves partial expression as a heterozygote.

More recently, it has been shown that the genetic interactions in Cystinuria are more complex than, though still quite comparable with, Harris' two phenotypes. Rosenberg, Downing, Durant & Segal (1966), studied the intestinal transport of the relevant amino acids and found evidence to suggest three genetically distinct diseases (see Table A-1, Appendix A). Type 1 comprised those Cystinurics who were unable to transport cystine and other basic amino acids in the gut, and whose parents were all completely recessive. Type 3 comprised those patients who transported some amino acids in the gut, and whose parents were all incompletely recessive. Type 2 was a much smaller group having some cystine but no lysine transport in the gut, and whose parents were also incompletely recessive.

Recently, individuals with a new genetic variant have been identified (Morin, Thompson, Jackson & Sass-Kortsak, 1971). These individuals, the "double heterozygotes", are heterozygotes for both the completely and incompletely recessive variant. Their amino acid excretion level is intermediate between the incompletely recessive heterozygote and the homozygous Cystinuric. They are therefore much less prone to form renal stones than are the homozygotes.

#### Incidence of Cystinuria

Reports on the incidence of Cystinuria in the general population, though not numerous, have varied among the different populations studied; and these variations may reflect true differences (see Table 1). However,

TABLE 1

Incidence of Heterozygous and Homozygous Cystinuria in the General Population

Author	Loc- al	Method of Detection	Number of Subjects	Incidence	
				Heterozygotes	Homozygotes
Lewis (1932)	U.S.	Nitroprusside,	10,534	approx. 1/600	-
Bostrum et al. (1959)	Swe- den	Nitroprusside & Paper Chromato.	7,793	approx 1/300	1/2,600.
Crawhall et al. (1966)	Eng.	Nitroprusside etc.	1,060	approx. 1/100	1/40,000 (est)
Levy et al. (1970)	U.S.	Nitroprusside & Paper Chromato.	117,000	-	1/23,000
Scriver et al. (1970)	Can- da	Nitroprusside & Paper Chromato.	4,714	approx. 1/50	1/10,000 (est)

Note.-- It is assumed that the ratio of incompletely recessive to completely recessive is 1 : 1 . Where original data were available this correction was made.



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results have also varied as a function of differences in method of detection and as a result of some confusion in defining homozygous or classical Cystinuria.

Lewis (1932), using non-specific tests which in addition to the target sample included many false positives, heterozygotes for Cystinuria and others, estimated the incidence of this entire group at 1/600. Bostrum and Tottie (1959), in a comprehensive study undertaken in Sweden, sampled 7,793 subjects using a more specific technique. They found three Cystinurics (1/2,600) and 14 incompletely recessive heterozygotes (completely recessive heterozygotes, since they have normal urine, would not be detected). Crawhall, Sanders & Thompson (1966) studied 1,060 patients in a general hospital, in England, and found that approximately 1/200 incompletely recessive heterozygotes were identified by the non-specific nitroprusside test. Since Crawhall had noted that the incidence of completely recessive Cystinuria is at least as great as that of the incompletely recessive type, then the nitroprusside test would detect only about 1/2 of heterozygotes. Through probabilistic inference (Hardy-Weinberg law) it was then concluded that the incidence of homozygous Cystinuria was about 1/40,000. Levy et al. (1970), examined specimens from 117,000 postnatal infants and discovered homozygous Cystinuria in five cases (1/23,000). In a more recent study of 4,714 general hospital patients, Scriver, Whelan, Clow & Dallaire (1970), found 43 or about 1/100 incompletely recessive heterozygotes. Through the Hardy-Weinberg law they inferred an expected frequency of homozygotes, in the general population, at about 1/10,000. From these studies it is conceivable that the true incidence of homozygous Cystinuria in the general population, in North America and England at any rate, is less than and probably considerably

less than 1/10,000.

### Treatment

There is no cure for this disorder. Treatment is designed to check the formation of stones and/or dissolve stones already present. It consists of drinking large quantities of fluids, and of ingesting sodium bicarbonate and D-penicillamine.

Drinking large quantities of fluids maintains a low concentration of cystine in the urine and, therefore, prevents precipitation and subsequent formation of stones. The Cystinuric is obliged to wake and drink during the night, each night, to maintain this high urine fluid level. Most Cystinurics are able to prevent stone formation by this day-night fluid intake regime, however, when stones are already present or form frequently the other treatments become necessary.

Sodium bicarbonate helps dissolve Cystine stones, however, the necessary ingestion of a large number of these tablets is unpleasant. D-penicillamine is a drug which combines with cystine in the kidney to form a product dissolving easily in urine and so reducing the amount of free cystine passed. However, this drug is used less frequently due to occasional unpleasant side-effects.

If the above treatment proves insufficient, as it does from time to time among some Cystinurics, then stones must be removed through surgery.

### Further Complications in Cystinuria

In addition to renal stones and expected concomitants, many other clinical features have been found coincident with the disorder. A recent review (van Sande, Terheggen, Clara, Leroy, & Lowenthal, 1971), listed a

variety of neurological and more general disorders which have been found in association with Cystinuria, for example, Down's Syndrome (e.g., Tanguay, 1966), Epilepsy (e.g., Herman & Lee, 1935), various congenital malformations (e.g., Pruzanski, 1966), haemophilia (Dent & Harris, 1951), dwarfism (Fleming, Avery, Morgan, & Cone, 1963), and others. However, there has been no evidence to suggest that the above associations are other than coincidental. Bostrum & Hambreas (1964), in their statistical survey, reported that the disease is apparently more serious in men than in women. In those cases with frank renal failure, the average age at death was 37.3 years for men and 53.8 years for women. Collis, Levi & Milne (1963) found that the mean height of 44 Cystinurics, in Britain, was 2.5 cm. below the published general population mean, a significant difference. They suggested that this might be related to the Cystinurics' decreased absorption of amino acids. Of the clinical features which have been found coincident with Cystinuria, the association with mental retardation or other mental disorder has recently received most attention.

There is a small but growing number of studies which suggest that homozygous Cystinuria is significantly more prevalent among patients in mental institutions, than in the general population (see Table 2). Berry (1962) noted that in several metabolic diseases known to be associated with mental retardation (e.g., Phenylketonuria), characteristic substances are excreted in the urine. She obtained urine samples from 700 mental retardates, from 60 participating homes and agencies in Ohio, and found 3 with phenylketonuria, 3 with glycosuria, 1 with cystinuria (1/700), and 9 with gross aminoaciduria. Visakorpi & Hyske (1960) examined 177 retarded subjects and found one Cystinuric who was also an epileptic, while Efron (1965) found two Cystinurics in a sample of 2,000 patients from an institution for the retarded in

TABLE 2

Incidence of Homozygous Cystinuria Among Those in Mental Institutions

Author	Number of Subjects	Incidence of Homozygotes
Visakorpi & Hyrske (1960)	700	1/700
Berry (1962)	177	1/177
Carson & Neill (1962)	2,081	1/2081
Efron (1965)	2,000	1/1,000
Scriver et al. (1970)	1,400	1/700
Wadman & Van Sprang (1971)	4,000	1/1,333

Massachusetts. Carson & Neill (1962), in Ireland, discovered eight cases of Cystinuria in a sample of 2,081 mentally retarded children. Striver et al. (1970), in Montreal, obtained urine samples from 1,400 patients at a psychiatric hospital and found two previously undiagnosed Cystinurics. One, a Chinese with paranoid schizophrenia and under psychiatric care since 1964, was born in 1930 and was 38 years old at the time of the incidental discovery of Cystinuria. The other patient, white, born in 1951, was admitted to hospital at the age of 17 years with latent schizophrenia and a pathological personality. More recently, Wadman and van Sprang (1971) discovered three Cystinurics among a sample of 4,000 mental retardates.

From the above studies, the incidence of Cystinuria in the population of those in mental institutions seems to be about 1/1,000 (compared to the estimated frequency in the general population of less than 1/10,000). In other words, Cystinuria appears to be at least ten times as prevalent among those in mental institutions as in the general population. Reasons for this have been suggested. As reviewed earlier, it has been demonstrated that the amino acid transport systems of the brain closely resemble those of the kidney (Blasberg & Lajtha, 1965). It is therefore possible that a disorder, e.g., Cystinuria, impairing net renal cellular uptake of lysine, an essential amino acid, might have parallel effects in the brain and thus lead to impaired brain function and/or impaired development of cognitive skills. Van Sande et al. (1971), suggest that the observed association between Cystinuria and mental disorder "... could be due either to a biochemical defect involving both brain and kidney or both the renal lesions and the amino aciduria could be 'secondary rather than primary manifestations of the metabolic error.'"

Other renal transport aminoacidurias, Hartnup Disease and Methionine malabsorption, have already been found to be associated with mental retardation (Efron, 1965). Hartnup's Disease has, in addition, been associated with retarded growth.

The present study sought to enquire into a possible relationship between Cystinuria and intellectual impairment. In addition, the personality characteristics of Cystinurics were explored.

#### Cystinuria and Intellectual Impairment

It is probable that the normal distribution of intelligence is largely determined by many genes, each having minor effect (e.g., Carter, 1962). However all that has been found to-date are a small number of genes, each with a major influence on intelligence. Since it seems that a small yet disproportionately large number of Cystinurics suffer substantial intellectual deficits, then perhaps Cystinuria is one of these hypothesized factors of minor influence. If this is so, then it may be that each Cystinuric is minimally impaired in his intellectual development and/or functioning, such that the sampling distribution of I.Q. for Cystinurics continues to approximate the normal distribution but is shifted downwards and, therefore, includes more mental retardates (e.g., see Figure 1). For example, the expected frequency of "mental defectives" (I.Q. below 70) in the general population is 2.2% (Wechsler, 1949, 1955). However, if the mean of an approximately normal distribution of I.Q. scores for Cystinurics was 95, or 1/3 of a standard deviation below the general population mean of 100, then approximately 4.9% of Cystinurics would be "mentally defective".

Alternatively, it may be that most Cystinurics are not mentally abnormal but that perhaps only one type of Cystinuria leads to

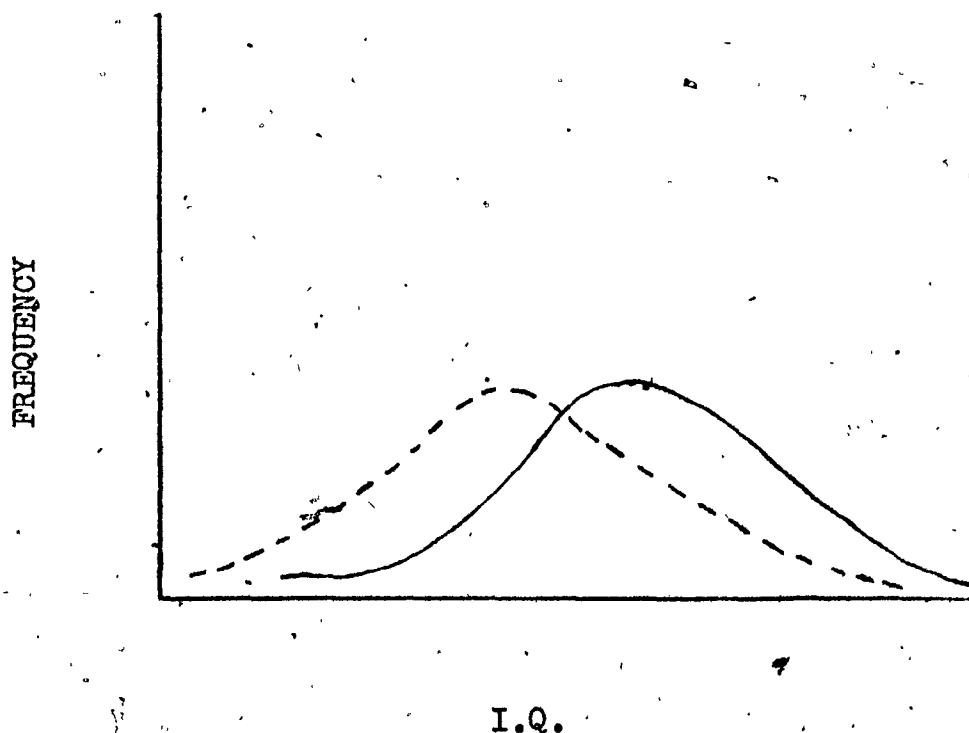


Figure 1. Sampling distribution of I.Q. in the normal population, mean of 100, (solid line) compared to a hypothetical sampling distribution of I.Q. for Cystinurics, mean  $< 100$  (broken line).

significant intellectual impairment. It is also possible that mental retardation does not occur in the absence of another genetic variant co-existing with Cystinuria. A disproportionately large number of mental retardates among an otherwise intellectually normal population of Cystinurics would give rise to a non-normal sampling distribution of I.Q.'s with an over-representation at the lower end (e.g., see Figure 2).

Based on considerations reviewed above it was hypothesized that the Cystinuric has a greater likelihood of intellectual impairment than has a non-cystinuric, all other factors being equal.

#### Cystinuria and Personality Differences

Internal versus external control of reinforcement. Personality is often studied by considering an individual's traits or stable characteristics which are then used as descriptive and predictive variables. One of these general personality characteristics is the internal-external (I-E) control dimension as derived from social learning theory (Rotter, 1954). According to this hypothesis there is an internal-external continuum. At one extreme of this continuum (Rotter, 1966) are those individuals who, on the basis of past experience, learning and generalization, believe that rewards and punishments have an external locus of control and are therefore the result of chance or luck or other factors beyond their control. At the other end of the continuum are those, the internals, who feel that their own actions, capacities, or traits are responsible for the things which happen to them (Hersch & Scheibe, 1967). Of course, most individuals fall between these two extremes.

The above theory has been subjected to much experimentation, primarily through the use of I-E scales and their variants (e.g.,



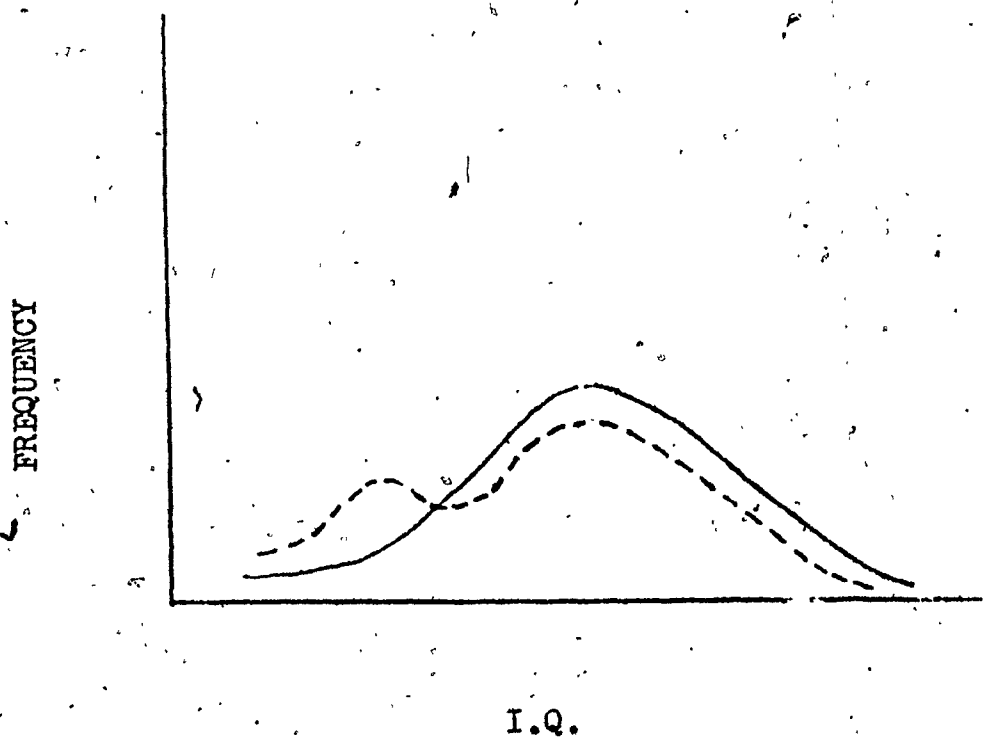


Figure 2. Sampling distribution of I.Q. in the normal population, mean of 100, (solid line) compared to one possible hypothetical sampling distribution of I.Q. for Cystinurics (broken line), where there is a disproportionately large frequency at the lower end.

Rotter, Seeman, & Liverant, 1962; Bialer, 1961; Crandall, Katkovsky & Crandall, 1965). This has subsequently led to the view that this concept or construct is useful in the study of personality (Rotter, 1966; Lefcourt, 1966; Joe, 1971).

Internality-externality, as a general personality trait or characteristic, has often been found, through experimentation, to be a significantly effective predictor of individual differences in reinforcement sensitivity, in attitudes, and in social behaviour. For example, Seeman & Evans (1962) worked with patients in a tuberculosis hospital. They found, as expected, that internals (defined in most studies as those scoring below the median for the group under investigation) were more inclined to seek out information about their condition and were generally less satisfied with feedback from doctors and nurses. Seeman (1964) found that internals joined more and were more active in unions and had a greater interest in the knowledge of political events. Hersch and Scheibe (1967) concluded that increased externality related positively to measures of maladjustment, in addition, internals described themselves as more active, striving, achieving, powerful, independent, and effective. Feather (1967) noted a significant tendency for externally scoring subjects, of both sexes, to report more debilitating anxiety and more neurotic symptoms. Phares, Ritchie and Davis (1968) concluded that internals were more willing to remedy personality problems, while MacDonald (1970) noticed that internals were more inclined to practice some form of birth control than were externals. It is also noteworthy, particularly for this study, that correlations between I-E and I.Q. have been on the whole very low (Rotter, 1966).

Rotter (1966), in a review, concluded that I-E is a useful and

valid concept and that based on previous studies internals are more likely, to be alert to aspects of the environment which might later benefit them, to take steps to improve their environmental condition, to place greater value on skill or achievement reinforcements and be generally more concerned with their ability or inability to succeed, and be more likely to resist subtle attempts to influence them.

Joe (1971), in a review of the more recent literature on the I-E dimension, has largely reaffirmed Rotter's conclusions.

#### Cystinuria and internal versus external control of reinforcement.

Cystinuria is a disorder which does not often incapacitate the Cystinuric, in any substantial manner throughout his life. However, once the disease is identified, there is a continuous lifelong awareness and inconvenience associated with treatment. In prevention of stone formation the Cystinuric must awaken during the night, each night, to drink water. If a stone is formed, the unpleasantness associated with the sodium bicarbonate or the occasional side-effects due to the D-penicillamine, in addition to the clinical feature of pain, add to this inconvenience. Furthermore, surgical intervention is not infrequent, with many Cystinurics periodically undergoing operations.

Through the frame of reference established in social learning theory, being a Cystinuric might be regarded as a major aversive stimulus or "punishment" which is not contingent on previous behavior but occurs essentially by chance. The Cystinuric has no control over whether or not he is a Cystinuric. It is not inconceivable, therefore, that such a situation, aside from the specific behavioral adjustments necessitated, might have an effect on the individual's outlook on and style

of interaction with the environment. This may be especially true if the disorder is identified early in life, as is usual.

One's perception of locus of control of reinforcement is determined by previous experiences. Since having to live with Cystinuria must be regarded as a particularly significant set of experiences, then it might be suggested that the Cystinuric's felt locus of control will tend to shift somewhat toward the external end of the I-E continuum.

Cystinuria and general personality differences. At least one study (Scriver et al., 1970) has suggested that Cystinurics are more prone to psychopathology, than are others. However, due to the dearth of information on personality characteristics of Cystinurics as a group; no firm hypotheses were possible. In fact, this part of the study was seen more as an exploration into the personality and general life-adjustment of those with the disease. It was hypothesized that Cystinurics, because of organic brain factors and/or because of personality characteristics resulting from knowledge of and necessity of dealing with the disease and/or because of other's reactions to the disorder, are psychologically different from the non-cystinuric. It was further hypothesized that these differences would tend to be in directions suggesting a poorer general life-adjustment among Cystinurics.

#### Summary of Hypotheses

It was hypothesized that the Cystinuric has a greater likelihood of intellectual impairment than his non-cystinuric sibling. In this study the non-cystinuric sibling served as a member of the control group, and this group loosely represented the normal population. Cystinurics should therefore, on the average, have a lower I.Q. than their non-cystinuric siblings.

It was hypothesized that the Cystinuric would tend to be, on the average, more external on the I-E continuum than his non-cystinuric sibling.

It was hypothesized that an objective test of personality and a structured interview would reflect some systematic differences between Cystinurics and their non-cystinuric siblings, and that Cystinurics would exhibit a poorer general life-adjustment.

## Method

Subjects

Employing the matched pairs design whenever possible, a total of 38 Caucasian subjects ranging in age from 7 years 11½ months to 48 years, and including 26 Cystinurics and 12 controls (non-cystinuric sibs), were obtained from the general population in several centres in Canada and in the United States (see Tables B-1, B-2, Appendix B). Each subject's first language was English. All of the Cystinurics and all but one of the controls were authenticated through hospital records (see Table B-3, Appendix B). The remaining control is 36 years old, male, and has never had kidney problems. If he were Cystinuric, there is a greater than 90% chance he would have formed a stone by now (Crawhall, 1973). Therefore, he was included in the sample as a control. There were, consequently, 12 Cystinuric-non-cystinuric sibling pairs, matched as closely as possible for age, education, and sex.

Materials

Intellectual assessment measures. Each subject was administered an individual test of intelligence, either the Wechsler Adult Intelligence Scale, (WAIS), (Wechsler, 1955), or the Wechsler Intelligence Scale for Children, (WISC), (Wechsler, 1949), for those under 16 years of age.

The WAIS and the WISC each consists of 11 subtests (see Appendices C & D for notes on subtests and sample questions). Six of these are grouped into a Verbal scale while all 11 subtests, Verbal and Performance scales, are combined to make the Full Scale I.Q. Both the WAIS and the WISC are well standardized, highly reliable, and quite valid (Wechsler,

1949; 1955).

Each of the 11 WAIS subtests measures a cognitive ability which is virtually identical to the ability measured by one of the WISC subtests. Verbal, Performance, and Full Scale I.Q.'s are age corrected and expressed as standard scores, therefore, I.Q.'s earned on the WAIS are quite comparable with those earned on the WISC. Similarly, WISC subtest standard scores and age-corrected WAIS subtest standard scores are also quite comparable.

Personality assessment measure, Internality-Externality.

Each adult subject completed Rotter's I-E scale (Rotter, 1966), which consists of 23 scored items and 6 fillers (see Appendix E). The scale is of the forced choice type, and the two statements in each pair, one external and one internal, have been judged to have equivalent social acceptance value (Rotter, 1966). Item analysis and factor analysis show reasonably high internal consistency for an additive scale (Rotter, 1966). Test-retest reliability is satisfactory, .49-.83, and the scale correlates well with other methods of assessing the same variable such as questionnaire techniques, Likert scales, interview assessments, and a story completion technique (Rotter, 1966).

The above I-E scale is designed for adults. A number of related scales have been designed expressly for children, but none of these are directly comparable to Rotter's scale. However, a variant of Bialer's Locus of Control Scale (Bialer, 1961), the Perceived Internal Locus of Control Scale (Gochman, 1971) is the most similar to the I-E and, therefore, was used in this study.

The Perceived Internal Locus of Control Scale, or PILC, (Gochman, 1971), is a 23 item scale with a forced choice Yes or No

response required for each item (see Appendix F). Bialer's scale has been reported to have reasonable internal consistency (Lefcourt, 1966).

The I-E scale and the PILC are consistent in that they attempt to measure a general personality dimension of internal versus external locus of control. Both scales contain test items in a number of motivational and behavioral areas such as affiliation, dominance, achievement, and dependency. However, scores on these tests are not sufficiently comparable to permit grouping for statistical analyses. Therefore, it was necessary to be flexible in administration so that, whenever possible, both siblings in each matched pair completed either the I-E scale or the PILC. This flexibility was possible as either scale is appropriate for those who are 15-17 years of age.

In each of the scales the score equals the total number of external items chosen. Therefore, theoretically, the higher the score the more external is the individual.

Personality assessment measures, the 16 PF, the HSPQ, and the CPQ.

Each subject completed a general, objective, personality inventory. Three similar and highly comparable self-report inventories were used. These were the Cattell Sixteen Personality Factor Questionnaire, (16 PF), (Cattell, Eber & Tatsouka, 1969), for those 16 years of age and older; the Jr.-Sr. High School Personality Questionnaire, (HSPQ), (Cattell, R.B., & Cattell, M.D., 1969), for those 12-18 years of age; and the IPAT Children's Personality Questionnaire, (CPQ), (Porter & Cattell, 1963), for those 8-12 years of age (see Appendix G, H & I, respectively, for test forms). All of these scales are well standardized, reliable, and valid (Cattell, R.B., & Cattell, M.D., 1969; Cattell, Eber & Tatsouka, 1970, Porter & Cattell, 1968).



Cattell's 16 PF test was constructed through factor-analytic studies and thus items are grouped into 16 homogeneous and independent clusters, each with a bipolar title (see Appendix J). In addition to these 16 independent clusters, or primary factors, there are seven secondary factors, each derived from an appropriately weighted combination of a number of the former.

The HSPQ and the CPQ are identical to the 16 PF in the method of test construction, in test design and administration, and in the manner in which scores are expressed. These questionnaires each yield 14 primary and 4 secondary factors (see Appendices K & L).

For all forms, raw scores are converted to stens through normative tables published by the test authors. These tables conform to a symmetrical sampling distribution with a mean of 5.5 and a standard deviation of two. In this study each individual's raw scores were converted to stens through the norm table most specific to that individual. Scores expressed as stens were then age-corrected.

There are 11 source and four secondary factors common among the above three scales (see Appendices J, K & L). Since these scales are so highly comparable, the stens for each of the above factors were grouped for statistical comparison without regard to the inventory on which they were achieved. Only these 15 factors were evaluated statistically.

Personality assessment measures, the structured interview.

Each subject participated in an orally administered and tape recorded structured interview designed to obtain life history data and more specifically information about development, school, personality, mood, and manner of interaction with others. Two interview forms were used,

one for those 16 years of age and over (see Appendix M), and a children's version (see Appendix N). The interview schedules were constructed from selected items taken directly from a number of published interview schedules (Burdock & Hardesty, 1968; Bennet & Orbach, 1946; Weider, Wolff, Brodman, Mittelmann, & Wechsler, 1948), or items taken and changed somewhat to suit the purposes of this study.

A scoring checklist, termed the Desirability Scale (see Appendix O), was largely constructed independently although many of the items were selected from the Structured Clinical Interview (Burdock & Hardesty, 1968). This scale represented an attempt to order the interview data in a systematic manner, with a view to statistical analysis. In this Desirability Scale the total score is indicative of the desirability of an individual's qualities, activities, and life-situation as presented by him (or her) in the interview. The more negative the score the less the desirability.

Validity and reliability of this scale are unknown. However, three measures of interscorer reliability were obtained from three raters. Two independent raters, naive to the purposes of the study, scored the Desirability Scale as they listened to the tape-recorded interview. Each rater evaluated 14 subjects in this manner. The interviewer, having filled out the Desirability Scale as soon as possible after the completion of the interview, served as a third rater. Some of the taped interviews were unavailable for scoring by the two naive raters. Therefore, to maintain consistency, the Desirability Scale scores involved in statistical comparisons of groups were exclusively those of the interviewer.

Procedure

Many individuals were involved in locating subjects and in obtaining their assistance. Cystinurics or the parents of child Cystinurics were contacted by their physician, by their physician's assistant, or directly by the researchers. Adult Cystinurics themselves arranged for the participation of non-cystinuric sibs.

Subjects were seen, at their convenience, in a variety of situations and under various testing conditions. Most subjects were seen in their homes, or at the homes of their sibs or parents (30 subjects). Of the others, two were seen in hospital, three at a University, two at a fraternity house, and one at a University dormitory. Due to logistical difficulties the experimenter was invariably aware of each subject's classification.

Subjects were aware that the purpose of the study was to obtain a better understanding of the effects of Cystinuria and that the researcher was, therefore, screening for possible intellectual and personality differences between the Cystinuric and his non-cystinuric sibling.

Each subject, seen only once and for about 2½ hours, was administered an individual I.Q. test, an I-E or PILC scale, and a general personality inventory, as well as participating in a structured interview. Assessment procedures were counterbalanced to control for possible carryover effects.

## Results

Matched Pair Comparisons for Twelve Cystinuric - Non-Cystinuric Sibling Pairs

Intellectual Evaluation. A summary of the statistical analyses on the mean I.Q. differences between Cystinurics and controls are presented in Table 3 (see Table P-1, Appendix P, for source data). The mean Performance I.Q.'s were virtually identical between groups ( $t = .02$ ,  $df = 11$ ,  $p > .05$ ). Though it was noted that the mean Full Scale and particularly that the mean Verbal Scale I.Q. scores of the Cystinurics exceeded those of the controls, these differences were insignificant ( $t = .95$ ,  $df = 11$ ,  $p > .05$  and  $t = 2.02$ ,  $df = 11$ ,  $p > .05$ , respectively).

The analysis of variance (2-within design) on standard scores for 10 of the 11 intelligence scale subtests is summarized in Table 4 (see Table P-2, Appendix P, for source data). The Digit Symbol or Coding subtest was omitted from this analysis as scores were unavailable for a number of the subjects. No significant differences were noted between the Cystinuric and the control group. Cystinurics and controls were not differentiated on the basis of combined scores across subtests for each of the two groups (Group effect) ( $F = .835$ ,  $df = 1$  and  $11$ ,  $p > .05$ ). In addition, the interaction between Group and Subtest was not significant ( $F = .4$ ,  $df = 9$  and  $99$ ,  $p > .05$ ). It was found, however, that the subtest scores differed significantly when they were analyzed independent of group classification ( $F = 5.92$ ,  $df = 9$  and  $99$ ,  $p < .05$ ).

In summary, the results consistently reflected no intellectual differences between the Cystinurics and the controls.

TABLE 3  
Significance of Mean I.Q. Differences  
Between Cystinurics and Controls

Intelligence Scale	Mean Deviation	t <sup>a</sup>
Verbal	5.08	2.02
Performance	.07	.02
Full	2.75	.95

a. two-tail test.

TABLE 4  
 ANALYSIS OF VARIANCE ON INTELLIGENCE  
 SCALE SUBTESTS

Source	df	MS	F
Groups (A)	1	9.6	.84
Subtests (B)	9	10.89	5.92*
Subjects (C)	11	61.5	
A X B	9	2.96	.4
A X C	11	11.5	
B X C	99	1.84	
A X B X C	99	7.4	

\*  $p < .05$

Personality Characteristics. The analysis of variance on standard scores for the 11 comparable and independent source factors, among the personality inventories, is summarized in Table 5 (see Table P-3, Appendix P, for source data). No significant differences were noted. Cystinurics and controls were not differentiated on the basis of combined scores for each group ( $F = 1.61$ ,  $df = 1$  and  $11$ ,  $p > .05$ ). Similarly, there was no interaction between Group and Factor; personality factor scores did not vary as a function of group classification ( $F = 1.51$ ,  $df = 10$  and  $110$ ,  $P > .05$ ).

Separate comparisons between the Cystinuric and the control group mean standard scores on each of the 11 source or primary factors are summarized in Table 6 (see Table P-4, Appendix P, for source data and Figure P-1, Appendix P, for a descriptive comparison of mean personality profiles for Cystinurics and controls). The only factor on which Cystinurics and controls differed significantly was Factor E (Submissiveness-Dominance) ( $t = 2.2$ ,  $df = 11$ ,  $p < .05$ ), with the affected subjects more dominant than their non-cystinuric sibs. There was a trend toward significance on Factor C (Lower-Higher Ego Strength) ( $t = 2.12$ ,  $df = 11$ ,  $p < .10$ ), cystinurics tending to exhibit less ego strength than the controls. There was also a trend toward significance on Factor I (Tough minded-Tender minded) ( $t = 2.15$ ,  $df = 11$ ,  $p < .10$ ), with Cystinurics tending to be more tough-minded.

The analysis of variance on standard scores for the four secondary factors common among the personality inventories is summarized in Table 7 (see Table P-4, P-5 and Figure P-1, Appendix P, for source data). No significant differences were found. There was no overall Cystinuric - non-cystinuric group difference ( $F = .54$ ,  $df = 1$

TABLE 5  
 ANALYSIS OF VARIANCE ON PERSONALITY  
 SOURCE FACTORS

Source	df	MS	F
Groups (A)	1	4.9	1.61
Source factors (B)	10	6.01	1.16
Subjects (C)	11	3.39	
A X B	10	5.01	1.51
A X C	11	3.05	
B X C	110	5.18	
A X B X C	110	3.32	



TABLE 6  
 Significance of Mean Standard Score  
 Differences Between Cystinurics and Controls  
 for each Primary Personality Factor

Personality Factor Code	Mean Difference	t <sup>a</sup>
A	1.07	1.46
B	.33	.43
C	1.11	2.12 †
E	1.35	2.22 *
F	.33	.36
G	1.0	1.23
H	.4	.5
I	1.33	2.15 †
O	.7	1.15
Q <sub>3</sub>	1.15	1.49
Q <sub>4</sub>	.3	.50

<sup>a</sup> Two-tail test

† p < .10

\* p < .05

TABLE 7  
ANALYSIS OF VARIANCE ON SECONDARY  
PERSONALITY FACTORS

Source	df	MS	F
Groups (A)	1	1.9	.54
Secondary factors(B)	3	4.13	2.18
Subjects (C)	11	3.23	
A X B	3	2.27	1.91
A X C	11	3.53	
B X C	33	1.95	
A X B X C	33	1.19	

and 11,  $p > .05$ ), and there was no interaction between Group and Personality Factor ( $F = 1.91$ ,  $df = 3$  and  $33$ ,  $p > .05$ ).

The analyses of scores on measures of internality-externality were undertaken separately for children (PILC scale) and for adults (I-E scale). The median external scores for five child Cystinurics and their controls were 10 and 10.25 respectively. The median external scores for six adult Cystinurics and their controls were 10 and 9.75 respectively. The Sign Test applied to the PILC scale scores yielded no significant difference between Cystinurics and controls ( $N = 5$ ,  $X = 2$ ,  $p = .50$ ). Since no differences were observed in two of the six matched pairs, on the I-E scale, the sample size was reduced to below numbers amenable to statistical analysis. In addition, the four remaining matched pairs failed to suggest any trends in the data.

Inter-scorer reliabilities for the Desirability Scale, obtained among three raters, are summarized in Table P-6, Appendix P. Comparisons indicate that the closest inter-scorer agreement was obtained by the two independent and naive raters. Spearman Rank Correlation co-efficients were all highly significant and moderately high for a scale of this type ( $r_s = .794$ ,  $.691$  and  $.646$ , all at  $p < .01$ ).

The median Desirability Scale score for nine Cystinurics was -7 and the median for their controls was -4.33. The Sign Test analysis of the differences between Cystinurics and controls, on the Desirability Scale, reflects no significant difference ( $N = 8$ ,  $X = 3$ ,  $p = .363$ ).

In summary, the only significant personality difference found between Cystinurics and controls was that the former scored higher on Factor E (Submissiveness-Dominance) of the personality inventory.

Cystinurics were found to be more dominant than their non-cystinuric siblings.

Additional Results: A Comparison of the Total Sample of Twenty-Six Cystinurics with Normative Standards

The Cystinuric group mean Full Scale I.Q. of 106.25 (SD = 15.56) exceeded the established normative mean of 100 (SD = 15) to a significant degree ( $Z = 2.12$ ,  $df = 25$ ,  $p < .05$ ):

The Cystinuric group mean stens, for each of the 11 personality source factors, were compared statistically with the constant normative group mean of 5.5 and standard deviation of two (see Table P-7, Appendix P, for source data and Figure P-2, Appendix P, for a descriptive comparison of the sample group means with the normative mean). A significant difference was found only on Factor G (Expedient-Conscientious) ( $Z = 2.17$ ,  $df = 25$ ,  $p < .05$ ), with the Cystinurics scoring as more expedient.

Fifteen adult Cystinurics obtained a mean of 10.67 (SD = 5.43) on the I-E scale, while 11 child Cystinurics achieved a mean external score of 8.2 (SD = 3.43) on the PILC.

## Discussion

### Data Relevant to the Intellectual Assessment

The results of the present study are inconsistent with expectations. It was hypothesized that Cystinurics would perform more poorly than their non-cystinuric siblings on a test of intelligence. Cystinurics did not perform more poorly on measures of Full Scale, Verbal, and Performance I.Q., and an analysis of variance on subtest scores failed to reveal any group differences. In addition, the mean Full Scale I.Q. for 26 Cystinurics was not below the normative mean of 100. In fact, the total sample of Cystinurics earned a mean I.Q. significantly above 100. This finding was not entirely unexpected since the mean educational level for 13 adult Cystinurics, age 16 years and over, is above-average at 12.6 years.

It was suggested earlier that Cystinuria may detract minimally from the development and/or from the expression of intellectual abilities. If this were true then Cystinuria should lead to minor and perhaps even to specific decrements in intellectual skills among virtually all those affected, and thereby give rise to an approximately normal I.Q. distribution with a mean less than 100 (e.g., Figure 1). The present findings reject this suggestion. It appears rather clear from the results that the genetic mutation for homozygous Cystinuria does not lead directly to observable intellectual deficit.

The results of the present study are interesting when compared with the few but consistent reports suggesting that Cystinurics are over-represented among the mentally retarded. It may be that these earlier studies are misleading, either due to biased sampling or to difficulties in ascertainment of Cystinuria, and that Cystinurics are

not really over-represented in mental institutions. Crawhall (1973), reviewed a report of a screening program for Cystinuria and other disorders in a mental institution (Scriver et al., 1970). He suggested that the two previously undiagnosed homozygous cystinurics (HC) discovered, there might be Cystinurics of a phenotypically very similar but genotypically different type, the double heterozygote (DH) genetic compound. Although Crawhall's speculation is questioned (the above two subjects were believed to be Type 3 Cystinurics) his suggestion remains interesting as a general issue. DH have only recently been differentiated from HC, it is therefore likely that previous estimates of the incidence of HC, when based on direct observation, reflect the combined incidence of HC and DH. The incidence of HC when estimated from the heterozygote frequency likely reflects a more accurate estimate of the prevalence of the disorder. The reports on the incidence of HC in the general population have been based both on probabilistic inference and on direct observation, while reports on the incidence of the disease in mental institutions are all based on direct observation. It is possible, therefore, that the reported over-representation of this disorder among the mentally retarded may be misleading, if the genotype is an important determinant of cerebral phenotype.

Alternatively, the present results are not directly comparable with and, therefore, not necessarily contradictory to earlier findings. In the earlier studies subjects were from mental institutions, while the present subjects were members of the general noninstitutionalized population, and were volunteers. In fact it may be that the present and earlier findings, together, tend to support the suggestion of a disproportionately large number of mental retardates among an otherwise intellectually normal population of Cystinurics (e.g., see Figure 2). There are a number of possible explanations which could account for this finding. There may be a phenotypic variant for Cystinuria which leads to mental abnormality. For example, perhaps completely recessive

Cystinurics (Type 1) may be more prone to intellectual impairment because they are nutritionally disadvantaged (completely recessives do not transport cystine, lysine and arginine in the gut). Alternatively, it may be that Cystinurics do not suffer intellectual impairment in the absence of certain genetic factors, other than Cystinuria.

It is suggested that a follow-up psychological investigation limited to the families in which Cystinuria has been associated with mental retardation would prove valuable. The ideal study would include an evaluation of at least three siblings in each family: the mentally retarded Cystinuric, any one or all of his affected siblings whether or not they are retarded, and any one or all of his non-cystinuric sibs. The nature of the relationships among these three groups would not only be useful in further explaining the significance of the present findings, but would certainly help to clarify the relationship between Cystinuria and mental retardation.

#### Data Relevant to Internality-Externality

The absence of differences between the affected and the control group on measures of Internality-Externality are inconsistent with the expectation that Cystinurics would tend to be more external. Therefore, the suggestion that Cystinuria affords sufficient chance negative reinforcement so as to lead the Cystinuric to become more external is rejected.

However, perhaps expecting Cystinurics to be more externally oriented has simplified the true nature of the effects of the disorder. Though an affected individual is unable to effect a cure, he is usually quite able to control the disorder. It is possible, therefore,

that a tendency to become more external is counteracted through an individual's ability to control the effects of the Cystinuria. Under these circumstances no differences would be observed between Cystinurics and controls on the I-E, or on a related scale. Therefore other measures, besides I-E scales, would be required to test this hypothesis.

Additional findings of the mean external scores for 15 adult and for 11 child Cystinurics seem relatively consistent with mean scores obtained in other studies (e.g., Franklin, 1963; Ladwig, 1963). However, the small sample size and the absence of an appropriate normative group mean with which to compare the present findings prevents any firm statement based on these results.

#### Data Relevant to the General Personality Assessment

The findings of the general personality assessment were virtually all inconsistent with expectations. It was felt that Cystinurics would exhibit personality differences, when compared with their non-cystinuric sibs, and would do so in directions suggesting that they are less well-adjusted. Analysis of variance on 11 source and then on 4 secondary personality factors revealed no differences between the groups. Cystinurics and controls were not differentiated through the Desirability Scale. The only significant difference found among 11 separate factor-comparisons between matched pairs was on Factor E (Submissiveness-Dominance), with Cystinurics more dominant as a group than their controls. In addition, the only significant difference found among 11 separate factor comparisons in the overall Cystinuric sample was on Factor G (Expedient-Conscientious). The affected group was found to be more expedient. Among the many statistical analyses, which were performed, one or two significant findings are to be expected on the basis of chance.



Therefore, the hypothesis that Cystinurics possess different and less desirable personality characteristics and that they are less well adjusted was rejected.

The above conclusion applies only to members of the general population. As suggested earlier, a follow-up psychological investigation of families in which Cystinuria and mental retardation or other mental difficulties have been found to coincide, would be useful. However, since a personality assessment of a retarded individual is of dubious validity, the personality evaluation would necessarily be limited to the unretarded siblings in each family.

#### Limitations of the Study

The difficulty in obtaining subjects led to certain relaxations in controls which would otherwise not have been advisable. There were only 12 Cystinuric - non-cystinuric sibling pairs. Though matching was excellent for age and education, subjects were poorly matched for sex, and for degree of complications from Cystinuria. There were eight cross sex pairs, four of which consisted of a male Cystinuric with a female control. The inconsistency in the degree of complication arising from the Cystinuria, is significant, since suggestions of personality differences were based, partly, on the severity of the disorder.

To permit data analyses over a large age range, the standard scores achieved on the WISC and on the WAIS intelligence scales were grouped together. This was also done among the 16PF, HSPQ, and CPQ personality inventories. Although standard scores for the intelligence scales and among the personality inventories are highly comparable across forms, such comparisons are not as good nor as valid

as comparisons within any one scale or inventory. Despite the limitations of the study the rather consistent rejection of hypothesized psychological difficulties among Cystinurics strongly suggests that the present findings are valid.

## REFERENCES

- Bennet, G.K., & Orbach, C.E. Guidance summary form for use in vocational and educational counselling. New York: The Psychological Corporation, 1946.
- Berry, H.K. Detection of metabolic disorders among mentally retarded children by means of paper spot tests. American Journal of Mental Deficiency, 1962, 66, 555
- Bialer, I. Conceptualization of success and failure in mentally retarded and normal children. Journal of Personality, 1961, 29, 303-320.
- Blasberg, R. & Lajtha, A. Substrate specificity of steady-state amino acid transport in mouse brain slices. Archives of Biochemistry and Biophysics, 1965, 112, 361.
- Bostrum, H., & Hambreaus, L. Cystinuria in Sweden VII. Clinical, histopathological and socio-medical aspects of the disease. Acta Medica Scandinavica, 1964, Supp. 411, 1-61.
- Bostrum, H., & Tottie, K. Cystinuria in Sweden II. The incidence of homozygous cystinuria Swedish schoolchildren. Acta Paediatrica, 1959, 48, 345-352.
- Burdock, E.I., Hardesty, A.S. Structured Clinical Interview. New York: Springer Publishing, 1968
- Carson, N.A.J., Neill, D.W. Metabolic abnormalities detected in a survey of mentally backward individuals in Northern-Ireland. Archs. Dis. Childh., 1962, 37, 505.
- Carter, C.O., Human Heredity. Baltimore: Penguin Books Inc., 1962
- Cattell, R.B., & Cattell, M.D. The High School personality questionnaire. Champaign: IPAT, 1969a
- Cattell, R.B., & Cattell, M.D. Handbook for the jr.-sr. high school personality questionnaire. Champaign: IPAT, 1969b
- Cattell, R.B., Eber, H.W., & Tatsouka M.M. The sixteen personality factor questionnaire (16PF). Champaign: IPAT, 1969.
- Cattell, R.B., Eber, H.W. & Tatsouka, M.M. Handbook for the sixteen personality factor questionnaire. Champaign: IPAT, 1970.

- Collis, J.E., Levi, A.J., & Milne, M.D. Stature and nutrition in cystinuria and Hartnup disease. British Medical Journal, 1963, I, 590.
- Crandall, V.C., Katkovski, W., & Crandall, V.J. Children's beliefs in their own control of reinforcements in intellectual-academic achievement situations. Child Development, 1965, 36, 91-109.
- Crawhall, J.C., Saunders, E.P., & Thompson, C.J. Heterozygotes for Cystinuria. Annals of Human Genetics, 1966, 29, 257.
- Crawhall, J.C., & Watts, R. Cystinuria. American Journal of Medicine, 1968, 45, 736-755.
- Crawhall, J.C. Cystinuria: Diagnosis and Treatment. Unpublished manuscript, Royal Victoria Hospital, Montreal, 1973.
- Dent, C.E., & Harris, H. The genetics of cystinuria. Annals of Eugenics, 1951, 16, 50.
- Dent, C.E., & Rose, G.A. Amino acid metabolism in cystinuria. Quarterly Journal of Medicine, 1951, 20, 205.
- Efron, M.L. Aminoaciduria. New England Journal of Medicine, 1965, 272, 1058-1067 and 1107-1113.
- Feather, N.T. Some personality correlates of external control. Australian Journal of Psychology, 1967, 19, 253-260.
- Fleming, W.H., Avery, G.B., Morgan, R.I., & Cone, T.E. Gastrointestinal malabsorption associated with cystinuria. Pediatrics, 1963, 32, 358.
- Garrod, A.E. Inborn errors of metabolism. Lancet, 1908, 2, 1, 73, 142, 214.
- Gochman, D.S. Some steps towards a psychological matrix for health behaviour. Canadian Journal of Behavioral Science, 1971, 3.
- Harris, H. Human biochemical genetics. Cambridge: At the University Press, 1959.
- Harris, H., Mittwoch, U., Robson, E.B., & Warren, F.L. Phenotypes and genotypes in cystinuria. Annals of Human Genetics, 1955, 20, 57.
- Harris, H. & Warren, F.L. Quantitative studies on the urinary cystine in patients with cystine stone formation and their relatives. Annals of Eugenics, 1953, 18, 125.

- Herman, L., & Lee, W.E. Cystine nephrolithiasis. Report of two cases. Annals of Surgery, 1935, 101, 746.
- Hersch, P.D., & Scheibe, K.E. Reliability and validity of internal-external control as a personality dimension. Journal of Consulting Psychology, 1965, 29, 184-186.
- Joe, V.C. Review of the internal-external control construct as a personality variable. Psychological Reports, 1971, 28, 619-640.
- Lefcourt, H.M. Internal versus external control of reinforcement: A review. Psychological Bulletin, 1966, 65, 206-220.
- Levy, H.L., Shih, V.E., Madigan, P.M., Karolkewicz, V., & MacCready, R.A. Results of a screening method for free amino acids in urine. Clinical Biochemistry, 1968, I, 208.
- Lewis, H.B. The occurrence of cystinuria in healthy young men and women. Annals of Internal Medicine, 1932, 6, 183.
- London, D.R., & Foley, T.H. Cystine metabolism in cystinuria. Clinical Science, 1965, 29, 129.
- MacDonald, A.P., Jr. Internal-external locus of control and the practice of birth control. Psychological Reports, 1970, 27, 206.
- Milne, M.D., Asatoor, A.M., Edwards, K., & Loughbridge, L. The intestinal absorption defect in cystinuria. Gut, 1961, 2, 323.
- Morin, C.L., Thompson, M.W., Jackson, S.H., & Sass-Kortsak, A. Biochemical and genetic studies in cystinuria: observations on double heterozygotes of genotype I/II. Journal of Clinical Investigations, 1971, 1961.
- Phares, E.J., Ritchie, D.E., & Davis, W.L. Internal-external control and reaction to threat. Journal of Personality and Social Psychology, 1968, 10, 402-405.
- Porter, R.B. & Cattell, R.B. The children's personality questionnaire. Champaign: IPAT, 1963.
- Porter, R.B., & Cattell, R.B. Handbook for the children's personality questionnaire. Champaign: IPAT, 1968.
- Pruzanski, W., Cystinuria and cystine urolithiasis in childhood. Acta Paediatrica. Stockholm, 1966, 55, 97.
- Rosenberg, L.E., Downing, S. Darant, J., and Segal, S. Cystinuria: biochemical evidence for three genetically distinct diseases. Journal of Clinical Investigations, 1966, 45, 365.

- Rotter, J.B. Social learning theory and clinical psychology. New York: Prentice-Hall, 1954.
- Rotter, J.B. Generalized expectancies for internal versus external control of reinforcement. Psychological Monographs, 1966, 80, (1, whole no. 609).
- Rotter, J.B., Seeman, M., & Liverant, S. Internal versus external control of reinforcement: a major variable in behavior theory. In N.F. Washburne (Ed.), Decisions, values and groups. Vol. 2, Permagon Press: London, 1962.
- Scriver, C.R., & Whelan, D.T. Cystinuria: concepts and new observations. In N.A.J. Carson & D.M. Raine (Eds.), Inherited disorders of sulfur metabolism, 8th symposium society for study of inborn errors of metabolism. London: Churchill Livingstone, 1971.
- Scriver, C.R., Whelan, D.T., Clow, C.L. & Dallaire, L. Cystinuria: increased prevalence in patients with mental disease. New England Journal of Medicine, 1970, 283, 783-786.
- Seeman, M. unpublished manuscript, 1964. After Rotter, J.B. Generalized expectancies for internal versus external control of reinforcement. Psychological Monographs, 1966, 80, (1, whole no. 609).
- Seeman, M., & Evans, J.W. Alienation and learning in a hospital setting. American Sociological Review, 1962, 27, 772-783.
- Tanguay, R.B. Cystinuria associated with mongolism and identification of an abnormal compound in urine. American Journal of Clinical Pathology, 1966, 46, 442.
- Thier, S., Fox, M., & Segal, S. Cystinuria: in vitro demonstration of an intestinal transport defect. Science, 1964, 143, 482.
- Van Sande, M., Terheggen, H.G., Clara, R., Leroy J.C., & Lowenthal, A. Lysine-cystine pattern associated with neurological disorders. In N.A.J. Carson & D.M. Raine (Eds.) Inherited disorders of sulfur metabolism, 8th symposium society for study of inborn errors of metabolism. London: Churchill & Livingstone, 1971.
- Visakorpi, J.K., & Hyrske, J. Urinary amino acids in mentally retarded patients. Annals paediatrica. Fenn, 1960, 6, 112.
- Wadman, S.K., & Van Sprang, F.J. Frequency of mental retardation and neurological disturbances in patients with cystinuria. In N.A.J. Carson & D.M. Raine (Eds.), Inherited disorders of sulfur metabolism 8th Symposium society for study of inborn errors of metabolism. London: Churchill & Livingstone, 1971.

Wechsler, D. Wechsler Intelligence Scale for Children.  
New York: The Psychological Corporation, 1949.

Wechsler, D. Wechsler adult intelligence scale. New York:  
The Psychological Corporation, 1955.

Weider, A., Wolff, H.G., Brodman, K., Mittelmann, B., &  
Wechsler, D. Cornell Index. New York: The Psychological  
Corporation, 1948.

Wollaston, W.H. On cystic oxide: a new species of urinary  
calculus. Transactions Royal Society of London, 1810, 100,  
223.

APPENDICES



## APPENDIX A

Comparison of Classifications of Cases of  
Cystinuria Proposed by Harris & Rosenberg  
and Their Colleagues

TABLE A-1

Comparison of Classifications of Cases of Cystinuria Proposed  
 By Harris & Rosenberg and Their Colleagues (Reprinted from an  
 article by J.C.Crawhall & R.Watts, in Nov. 1968, Amer. J. of Med.)

Harris et al. (1955)	Completely Recessive	Incompletely Recessive
Rosenberg et al. (1966)	Type 4	Type 2
Amino acid excretion in urine of heterozygotes	normal	cystine and lysine increased
In-vitro gut transport studies	no transport of cystine lysine or arginine	no transport of cystine and lysine, reduced transport of cystine but somewhat less than in normals.

APPENDIX B

Biographical and Medical Data on Subjects

TABLE B-1

## Information on Unmatched Cystinurics

Subject	Sex	Age	Education
1 <sup>a</sup>	M	17	9
2	M	25	15
3	M	12	7
4	M	14	8
5 <sup>b</sup>	M	13	9
6	F	11	6
7	M	15	9
8 <sup>c</sup>	F	18	12
9	M	19	13
10	M	15	10
11 <sup>d</sup>	F	28	16
12	M	24	16
13	M	15	7
14	F	27	16

Note.- Numbers refer to years.

a. Residing in Florida

b. Residing in Ontario

c. Residing in Kentucky

d. Residing in Quebec

TABLE B-2

Information on Matching of Cystinuric - Non-cystinuric Sibling Pairs

Pair Number	Cystinurics			Controls		
	Sex	Age	Education	Sex	Age	Education
1 <sup>a</sup>	F	18	12	M	15	9
2	M	32	16	F	35	16
3 <sup>b</sup>	M	46	11	F	47	11
4	M	38	11	F	32	9
5	M	11	6	F	8	2
6 <sup>c</sup>	M	22	9	M	13	6
7	F	12	6	M	8	3
8	F	14	7	M	10	4
9	F	15	9	F	16	10
10 <sup>s</sup>	M	48	8	M	36	9
11	F	13	8	M	9	3
12 <sup>d</sup>	M	11	5	M	7	1

a. Residing in Florida

b. Residing in Ontario

c. Residing in Kentucky

d. Residing in Quebec

TABLE B-3

Clinical Evidence for Identification of Subjects as Cystinuric or Non-Cystinuric ( Control )

Subject Number	Clinical Information				Phenotype of some Family Members
	Stone	Stone Analyzed	Urinalysis for Amino Acids	Phenotype	
1	Yes	Cystine	Yes	HC <sup>a</sup>	
2	Yes	Cystine	Yes	HC	
3	Yes	Cystine	Yes	HC	
4	-	-	Yes	HC	
5	Yes	-	Yes	HC Type 2	Father and mother Heterozygote Type 2
6	Yes	-	Yes	HC Type 2	Father and mother Heterozygote Type 2
7	Yes	Cystine	Yes	HC Type 2	Father compound Type 1/2, mother Heterozygote Type 2
8	Yes	Cystine	Yes	HC	
9	-	-	Yes	HC	
10	-	-	Yes	HC	

TABLE B-3 - continued

Subject Number	Clinical Information				
	Stone	Stone Analyzed	Urinalysis for Amino Acids	Phenotype	Phenotype of some Family Members
11	Yes	-	Yes	HC	
12	Yes	Cystine	Yes	HC	
13	Yes	-	Yes	HC Type 1 or 3	Father and mother Heterozygotes Type 1 or 3
14	Yes	-	Yes	HC	Brother HC
15	Yes	Cystine	Yes	HC	
16	Yes	Cystine	Yes	HC	
17	Yes	Cystine	Yes	HC Type 1	
18	-	-	Yes	Double Heterozygote Type 2, mother	Father Heterozygote Type 2, mother
19	Yes	-	Yes	HC Type 2	Father and mother Heterozygotes Type 2
20	Yes	Cystine	Yes	HC	

TABLE B-3 - continued

Subject Number	Clinical Information				Phenotype of some Family Members
	Stone	Stone Analyzed	Urinalysis for Amino Acids	Phenotype	
21	-	-	Yes	HC	
22	-	-	Yes	HC	
23	Yes	Cystine	Yes	HC	Father HC
24	Yes	Cystine	Yes	HC	Daughter HC
25	-	-	Yes	HC	Sister HC
26	Yes	-	Yes	HC Type 1	
27	-	-	Yes	Normal or Heterozyg.	Sister HC
28	-	-	Self Report		Brother HC
29	-	-	Yes	Normal or Heterozygote Type 1	Brother HC
30	-	-	Yes	Normal or Heterozygote Type 1 or 2	Brother Double Heterozygote Type 1/2



TABLE B-3 - continued

Subject Number	Clinical Information					Phenotype of some Family Members
	Stone	Stone Analyzed	Urinalysis for Amino Acids	Phenotype	Phenotype	
31	-	-	Yes	Heterozyg. Type 2	Heterozyg. Type 2	Brother HC Type 2
32	-	-	Yes	Normal or Heterozyg.	Normal or Heterozyg.	Parents Heteroz. Type 2 Three Sibs HC
33	-	-	Yes	Normal or Heterozyg.	Normal or Heterozyg.	Three Sibs HC
34	-	-	Yes	Normal or Heterozyg.	Normal or Heterozyg.	Three Sibs HC
35	-	-	Yes	Heterozyg.	Heterozyg.	Father and Sister HC
36	-	-	-	Heterozyg.	Heterozyg.	Brother HC
37	-	-	Yes	Normal or Heterozyg.	Normal or Heterozyg.	Two sisters HC
38	-	-	Yes	Normal or Heterozyg.	Normal or Heterozyg.	Father HC

TABLE B-3 - continued

Note.- Subjects numbered 1 - 14, inclusive, correspond to Cystinurics numbered 1 - 14 in Table B-1. Subjects numbered 15 - 26, inclusive, correspond to Cystinurics numbered 1 - 12 in Table B-2. Subjects 27 - 38, inclusive, correspond to controls numbered 1 - 12 in Table B-2.

<sup>a</sup> HC refers to Homozygous Cystinuria.

APPENDIX C

A Brief Description of the Subtests of the

Wechsler Adult Intelligence Scale

Subtest	Subtest Reflects	Sample Question or Description of Tasks
Information	Memory of distant events, education, cultural interests, and opportunities	What does rubber come from?
Comprehension	Judgement, interest in social conformity, awareness of reality problems	Why should people pay taxes?
Arithmetic	Ability to concentrate, mental alertness, capacity for organization of stimuli	How many oranges can you buy for 36 cents if one orange costs six cents?
Similarities	Verbal concept formation, capacity for verbal abstraction	In what way are an orange and a banana alike?
<del>Digit Span</del>	Immediate recall	I am going to say some numbers...when I am through say them right after me. 6 - 4 - 3 - 9
Vocabulary	Education, recall of old memories.	...what does winter mean?
Digit Symbol	Capacity for new learning, visual motor activity and efficiency of routine psycho-motor functioning	Matching from sample, matching symbols to digits.

Subtest

---

Subtest Reflects

---

Sample Question or Description of Tasks

**Picture Completion**  
Alertness, ability to differentiate essential from non-essential

Subject must indicate what important part is missing in each picture.

**Block Design**  
Visual-motor co-ordination and space orientation

Using blocks, a subject must construct designs identical to those presented on cards.

**Picture Arrangement**  
Sophistication, ability to anticipate initial acts or situations

Subject must rearrange pictures, in sequence, so that they tell a logical story.

**Object Assembly**  
Capacity for visual-motor organization combined with past memory or habit formation

Subject must put a puzzle together.

APPENDIX D

Information on Subtests of the Wechsler Intelligence Scale for Children

Subtest	Sample Questions and/or Task Description
Information	<p>From what animal do we get milk?</p> <p>How many pounds are there in a ton?</p>
General Comprehension	<p>What is the thing to do if a fellow (girl) much smaller than yourself starts to fight with you?</p> <p>Why is cotton fiber used in making cloth?</p>
Arithmetic	<p>Children are asked to count a row of blocks which have been placed in front of them (Subjects below 8, or older subjects suspected of mental deficiency begin with this question).</p> <p>A boy had 12 newspapers and sold 5. How many did he have left?</p>
Similarities	<p>Finish what I want to say. Lemons are sour but sugar is _____. (younger subjects, below age 8, begin with this question).</p> <p>In what way are Beer and Wine alike?</p>
Vocabulary	<p>Bicycle...what is a bicycle?</p> <p>Join....what does join mean?</p>
Coding (or Digit Symbol)	<p>A - Subjects must place appropriate symbols within given designs (for all those under 8).</p> <p>B - As in the adult version (for all those from 8 - 15, inclusive).</p>
Digit Span <sup>a</sup>	
Picture Completion <sup>a</sup>	
Block Design <sup>a</sup>	
Picture Arrangement <sup>a</sup>	

Object Assembly<sup>a</sup>

Note: A description of what each subtest reflects is not included above since this information is virtually identical to that in Appendix C.

a - as in the adult version, see Appendix C.

## APPENDIX E

## ROTTER'S INTERNAL-EXTERNAL SCALE

This is a questionnaire to find out the way in which certain important events in our society affect different people. Each item consists of a pair of alternatives lettered a or b. Please select the one statement of each pair (and only one) which you more strongly believe to be the case as far as you're concerned. Be sure to select the one you actually believe to be more true rather than the one you think you should choose or the one you would like to be true. This is a measure of personal belief: obviously there are no right or wrong answers.

In some instances you may discover that you believe both statements or neither one. In such cases, be sure to select the one you more strongly believe to be the case as far as you're concerned. Also try to respond to each item independently when making your choice; do not be influenced by your previous choices.

Note - \* Denotes internal response

- Numbers 1, 8, 14, 19, 24, & 27 are fillers.

## APPENDIX.E - Continued

- 1.a. Children get into trouble because their parents punish them too much.
- b. The trouble with most children nowadays is that their parents are too easy with them.
- 2.a. Many of the unhappy things in people's lives are partly due to bad luck.
- b.\* People's misfortunes result from the mistakes they make.
- 3.a.\* One of the major reasons why we have wars is because people don't take enough interest in politics.
- b. There will always be wars, no matter how hard people try to prevent them.
- 4.a.\* In the long run people get the respect they deserve in this world.
- b. Unfortunately, an individual's worth often passes unrecognized no matter how hard he tries.
- 5.a.\* The idea that teachers are unfair to students is nonsense.
- b. Most students don't realize the extent to which their grades are influenced by accidental happenings.
- 6.a. Without the right breaks one cannot be an effective leader.
- b.\* Capable people who fail to become leaders have not taken advantage of their opportunities.
- 7.a. No matter how hard you try some people just don't like you.
- b.\* People who can't get others to like them don't understand how to get along with others.



APPENDIX E - Continued

- 8.a. Heredity plays the major role in determining one's personality.
- b. It is one's experiences in life which determine what they're like.
- 9.a. I have often found that what is going to happen will happen.
- b.\* Trusting to fate has never turned out as well for me as making a decision to take a definite course of action.
- 10.a.\* In the case of the well prepared student there is rarely if ever such a thing as an unfair test.
- b. Many times exam questions tend to be so unrelated to course work that studying is really useless.
- 11.a.\* Becoming a success is a matter of hard work; luck has little or nothing to do with it.
- b. Getting a good job depends mainly on being in the right place at the right time.
- 12.a.\* The average citizen can have an influence in government decisions.
- b. This world is run by the few people in power, and there is not much the little guy can do about it.
- 13.a.\* When I make plans, I am almost certain I can make them work.
- b. It is not always wise to plan too far ahead because many things turn out to be a matter of good or bad fortune anyhow.
- 14.a. There are certain people who are just no good.
- b. There is some good in everybody.

## APPENDIX E - Continued

- 15.a.\* In my case getting what I want has little or nothing to do with luck.
- b. Many times we might just as well decide what to do by flipping a coin.
- 16.a. Who gets to be the boss often depends on who was lucky enough to be in the right place first.
- b.\* Getting people to do the right thing depends upon ability; luck has little or nothing to do with it.
- 17.a. As far as world affairs are concerned, most of us are the victims of forces we can neither understand, nor control.
- b.\* By taking an active part in political and social affairs the people can control world events.
- 18.a. Most people don't realize the extent to which their lives are controlled by accidental happenings.
- b.\* There is really no such thing as "luck".
- 19.a. One should always be willing to admit mistakes.
- b. It is usually best to cover up one's mistakes.
- 20.a. It is hard to know whether or not a person really likes you.
- b.\* How many friends you have depends upon how nice a person you are.
- 21.a. In the long run the bad things that happen to us are balanced by the good ones.
- b.\* Most misfortunes are the result of lack of ability, ignorance, laziness, or all three.

## APPENDIX E - Continued

- 22.a.\* With enough effort we can wipe out political corruption.
- b. It is difficult for people to have much control over the things politicians do in office.
- 23.a. Sometimes I can't understand how teachers arrive at the grades they give.
- b.\* There is a direct connection between how hard I study and the grades I get.
- 24.a. A good leader expects people to decide for themselves what they should do.
- b. A good leader makes it clear to everybody what their jobs are.
- 25.a. Many times I feel that I have little influence over the things that happen to me.
- b.\* It is impossible for me to believe that chance or luck plays an important role in my life.
- 26.a.\* People are lonely because they don't try to be friendly.
- b. There's not much use in trying too hard to please people. If they like you, they like you.
- 27.a. There is too much emphasis on athletics in high school.
- b. Team sports are an excellent way to build character.
- 28.a.\* What happens to me is my own doing.
- b. Sometimes I feel that I don't have enough control over the direction my life is taking.

## APPENDIX E - Continued

29.a. Most of the time I can't understand why politicians behave the way they do.

b.\* In the long run the people are responsible for bad government on a national as well as on a local level.

## APPENDIX F

PERCEIVED INTERNAL LOCUS OF CONTROL  
SCALE

In this form we are interested in learning how people feel about different things that happen to them or could happen to them.

Please answer Yes or No to each question depending on how you yourself feel about it, and not how you think other people would or should answer. It is important that you answer every question. Some of the questions are easy to answer others are harder but answer all of them as well as you can.

Remember, this is a form to see how you feel about different things so there are no right or wrong answers, this is not a test.

Note: \*Denotes internal response.

## APPENDIX F - Continued

Item	Circle Yes or No	
1. When somebody gets made at you, do you usually feel that you can do something about it?	Yes*	No
2. Is it possible for a person to be whatever he wants to be?	Yes	No*
3. Are people mean to you even if you do not do anything to make them mean?	Yes	No*
4. Do you usually ask someone first before you make up your mind about something	Yes	No*
5. Can you do anything about what is going to happen in the future?	Yes*	No
6. When people are good to you, is it usually because you did something to make them be good?	Yes*	No
7. Can you ever make other people do things you want them to do?	Yes*	No
8. Do you ever think that people your age can change things that are happening in the world?	Yes*	No
9. If someone was going to hit you, would you be <u>unable</u> to stop him?	Yes	No*
10. Can a person your age ever have his own way?	Yes*	No
11. Is it hard for you to know why some people do the things they do?	Yes	No*
12. When people are nice to you is it only because they are nice persons?	Yes	No*

## APPENDIX F - Continued

Item	Circle Yes or No	
13. If you do not want to be friends with someone, can he ever make you change your mind?	Yes	No*
14. Is it useless to think about what you will be when you get older?	Yes	No*
15. When someone gets mad at you, can you usually do something to make him your friend again?	Yes*	No
16. Is it true that people your age have nothing to say about where you are going to live?	Yes	No*
17. When you get in an argument, is it sometimes your fault?	Yes*	No
18. When nice things happen to you, is it only good luck?	Yes	No*
19. Do you often feel you get punished when you don't deserve it?	Yes	No*
20. Will people usually do things for you if you ask them?	Yes*	No
21. Do you believe a kid can usually be whatever he wants to be when he grows up?	Yes*	No
22. When bad things happen to you, could it be your fault that they happen?	Yes*	No
23. Can you ever know for sure why some people do certain things?	Yes*	No

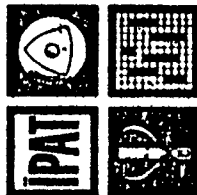
## APPENDIX G

The Sixteen Personality Factor Questionnaire:  
Test Form, Instructions to Subjects, and a  
Sample of Questions\*

\* The complete form includes 105 questions.



**16 PF**



**WHAT TO DO:** Inside this booklet are some questions to see what attitudes and interests you have. There are no "right" and "wrong" answers because everyone has the right to his own views. To be able to get the best advice from your results, you will want to answer them exactly and truly.

If a separate "Answer Sheet" has not been given to you, turn this booklet over and tear off the Answer Sheet on the back page.

Write your name and all other information asked for on the top line of the Answer Sheet.

First you should answer the four sample questions below so that you can see whether you need to ask anything before starting. Although you are to read the questions in this booklet, you must record your answers on the answer sheet (alongside the same number as in the booklet).

There are three possible answers to each question. Read the following examples and mark your answers at the top of your answer sheet where it says "Examples." Fill in the left-hand box if your answer choice is the "a" answer, in the middle box if your answer choice is the "b" answer, and in the right-hand box if you choose the "c" answer.

**EXAMPLES:**

1. I like to watch team games.  
a. yes,      b. occasionally,      c. no.
3. Money cannot bring happiness.  
a. yes (true),      b. in between,      c. no (false).

2. I prefer people who:
- a. are reserved,
  - b. (are) in between,
  - c. make friends quickly.

4. Woman is to child as cat is to:
- a. kitten, b. dog, c. boy.

In the last example there is a right answer—kitten. But there are very few such reasoning items.

Ask now if anything is not clear. The examiner will tell you in a moment to turn the page and start.

When you answer, keep these four points in mind:

1. You are asked not to spend time pondering. Give the first, natural answer as it comes to you. Of course, the questions are too short to give you all the particulars you would sometimes like to have. For instance, the above question asks you about "team games" and you might be fonder of football than basketball. But you are to reply "for the average game," or to strike an average in situations of the kind stated. Give the best answer you can at a rate not slower than five or six a minute. You should finish in a little more than half an hour.
2. Try not to fall back on the middle, "uncertain" answers except when the answer at either end is really impossible for you—perhaps once every four or five questions.
3. Be sure not to skip anything, but answer every question, somehow. Some may not apply to you very well, but give your best guess. Some may seem personal, but remember that the answer sheets are kept confidential and cannot be scored without a special stencil key. Answers to particular questions are not inspected.
4. Answer as honestly as possible what is true of you. Do not merely mark what seems "the right thing to say" to impress the examiner.

**DO NOT TURN PAGE UNTIL TOLD TO DO SO**

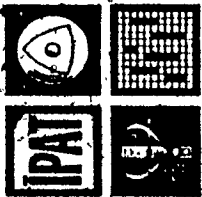
22. I am more annoyed by a person who:
- a. tells off-color jokes and embarrasses people,
  - b. uncertain,
  - c. is late for an appointment and inconveniences me.
23. I greatly enjoy inviting guests and amusing them.
- a. true, b. uncertain, c. false.
24. I feel that:
- a. some jobs just don't have to be done so carefully as others,
  - b. in between,
  - c. any job should be done thoroughly if you do it at all.
25. I have always had to fight against being too shy.
- a. yes, b. in between, c. no.
26. It would be more interesting to be:
- a. a bishop, b. uncertain, c. a colonel.
32. I like to join with people who show lively group enthusiasm.
- a. yes, b. in between, c. no.
33. I put my faith more in:
- a. insurance,
  - b. in between,
  - c. good fortune.
34. I can forget my worries and responsibilities whenever I need to.
- a. yes, b. sometimes, c. no.
- (End, column 2 on answer sheet.)
35. It's hard for me to admit it when I'm wrong.
- a. yes, b. sometimes, c. no.
36. In a factory it would be more interesting to be in charge of:
- a. machinery of keeping records,
  - b. in between,
  - c. talking to and hiring new people.
37. Which word does not belong with the other two?

27. If a neighbor cheats me in small things, I would rather humor him than show him up.  
a. yes, b. occasionally, c. no.
28. I like a friend who:  
a. is efficient and practical in his interests,  
b. in between,  
c. seriously thinks out his attitudes toward life.
29. It bothers me if I hear others expressing ideas that are contrary to those that I firmly believe.  
a. true, in between, c. false.
30. I am over-conscientious, worrying over my past acts or mistakes.  
a. yes, b. in between, c. no.
31. If I were good at both, I would rather:  
a. play chess,  
b. in between,  
c. go bowling.
32. a. cat, b. near, c. sun.
33. Minor distractions seem:  
a. to irritate me,  
b. in between,  
c. not to bother me at all.
34. I am quite happy to be waited on, at appropriate times, by personal servants.  
a. often, b. sometimes, c. never.
35. I would rather live in a town:  
a. artistically laid out, but relatively poor,  
b. uncertain,  
c. that is rough, prosperous, and booming.
36. People should insist more than they now do that moral laws be followed.  
a. yes, b. sometimes, c. no.
37. I have been told that, as a child, I was rather:  
a. quiet and kept to myself,  
b. in between,  
c. lively and always active.

## APPENDIX H

The Jr. - Sr. High School Personality  
Questionnaire: Test Form, Instructions to  
Subjects, and a Sample of Questions\*

\* The complete form includes 142 questions.



Jr.-Sr.

# HSPQ

FORM A

1968-69 Edition

**WHAT TO DO:** You have a Booklet and an Answer Sheet. Write your name, age, etc., on the Answer Sheet where it tells you to.

The Booklet before you has in it questions about your interests and your likes and dislikes. Although you are to read the questions in *this* Booklet, *you must put your answers on the Answer Sheet*, making sure that the number of your answer matches the number of the question in the Booklet.

First, we shall give you two examples so that you will know exactly what to do. After each of the questions there are three answers. Read the following examples and fill in the right boxes where it says Example 1 and Example 2, on the Answer Sheet, below your name. Fill in the left-hand box if your answer ~~choice~~ is the "a" answer, the middle box if your choice is the "b" answer, and the right-hand box if you choose the "c" answer.

**EXAMPLES:**

1. Which would you rather do:
  - a. visit a zoo,
  - b. uncertain,
  - c. go up in an airplane?
  
2. If you have a quarrel, do you make friends again quickly?
  - a. yes,
  - b. in between,
  - c. no.

As you see from these examples, there are usually no right or wrong answers, although sometimes a correct answer is expected. Each person is different and you should say only what is true for you. You can always find one answer that suits you a little better than the others, so never leave a question without marking one of the answers.

Inside you will find more questions like the ones above. When you are told to turn the page, begin with number 1 and go on until you finish all the questions. In answering them, please keep these four points in mind:

1. Answer the questions frankly and truthfully. There is no advantage in giving an untrue answer about yourself because you think it is the "right thing to say."
2. Answer the questions as quickly as you can. Don't spend too much time thinking about them. Give the first, natural answer that comes to you. Some questions may seem much like others, but no two are exactly alike so your answers will often be different too.
3. Use the middle answer only when it is absolutely impossible to decide on one of the other choices. In other words, the "a" or the "c" answer should be used most of the time.
4. Don't skip any questions. Sometimes a statement may not seem to apply to you, but answer every question, somehow.

If there is anything you don't understand, please ask your questions now. If you have no question now, but later on come across a word you don't know, ask the examiner then.

DO NOT TURN PAGE UNTIL TOLD TO DO SO

102. In talking with your classmates, do you dislike telling your most private feelings?  
a. yes, b. sometimes, c. no.
103. When you go into a new group, do you:  
a. quickly feel you know everyone,  
b. in between,  
c. take a long time to get to know people?
104. Look at these five words: *mostly, gladly, chiefly, mainly, highly*. The word that does not belong with the others is:  
a. mostly, b. gladly, c. highly.
105. Do you sometimes feel happy and sometimes feel depressed without real reason?  
a. yes, b. uncertain, c. no.
106. When people around you laugh and talk while you are listening to radio or TV:  
a. are you happy,  
b. in between,  
c. does it spoil things and annoy you?
107. If you accidentally say something odd in company, do you stay uncomfortable a long time and find it hard to forget?  
a. yes, b. perhaps, c. no.
113. How often do you go places or do things with a group of friends:  
a. very often, b. sometimes, c. hardly ever.
114. What kind of movie do you like best?  
a. musicals, b. uncertain, c. war stories.
115. Do you get in trouble more often by saying to a group that wants to do something:  
a. "Let's go!"  
b. uncertain,  
c. "I'd rather not join in"?
116. When you were growing up, did you expect the world to be:  
a. kinder and more considerate than it is,  
b. uncertain,  
c. tougher and harder than it is?
117. Do you find it easy to go up and introduce yourself to an important person?  
a. yes, b. perhaps, c. no.



118. Do you think that often a committee of your classmates takes more time and makes poorer decisions than one person would?  
a. yes, b. perhaps, c. no.

119. Do you feel you are doing pretty much what you should be doing in life?  
a. yes, b. uncertain, c. no.

120. Do you sometimes feel so mixed up that you don't know what you are doing?  
a. yes, b. perhaps, c. no.

121. When someone is disagreeing with you, do you:  
a. let him say all he has to say,  
b. uncertain,  
c. tend to interrupt before he finishes?

108. Which would you rather read about:  
a. how to win at basketball,  
b. uncertain,  
c. how to be nice to everyone?

109. Are you best thought of as a person who:  
a. thinks, b. in between, c. acts?

110. Do you spend most of your weekly allowance for fun. (instead of saving some for future needs)?  
a. yes, b. perhaps, c. no.

111. Do other people often get in your way?  
a. yes, b. in between, c. no.

112. How would you rate yourself?  
a. inclined to be moody,  
b. in between,  
c. not at all moody.

## APPENDIX I

The Children's Personality Questionnaire:  
Test Form, Instructions to Subjects, and  
a Sample of Questions\*

\* The complete form includes 140 questions,  
70 questions each for Parts A<sub>1</sub> and A<sub>2</sub>.



# CPQ, Form A (1963 Edition)

## Part A

### What You Do and What You Think

Print Your Name: First \_\_\_\_\_ Last \_\_\_\_\_

Your Age \_\_\_\_\_ Grade in School \_\_\_\_\_ Boy or Girl \_\_\_\_\_

Read each statement and mark an  on the side that fits you better. Some questions will not have the words just the way you want them but mark every one the best you can. You may ask for help if you don't know a word. Just raise your hand and the teacher will come to your desk. Do not work long on one question. Mark it and go right on to the next one. **MARK EVERY ONE.** Most of the questions have two boxes to choose from but other questions have three boxes. Always look at ALL the boxes and pick just one of them for your answer.

- 1. When visiting a new building do you like to have someone show you around  or  do you like to find your own way
- 2. When a child laughs at you do you feel badly  or  do you laugh too
- 3. Do you think you could do well at almost anything  or  just a few things
- 4. In a game on the playground, do you stand around  or  run a lot
- 5. Does your mother think you are too lively and restless  or  quiet and calm
- 6. Do you feel nervous at school  or  are you happy

- 7. Do you work slowly  or  quickly
- 8. In your group is someone else the leader  or  are you the leader
- 9. Do you have many friends  or  just a few good friends
- 10. Do you think you smile a great deal  or  do not smile much

**GO RIGHT ON TO THE NEXT PAGE.**

DO NOT WRITE BELOW THIS LINE.

FACTOR	A <sub>1</sub>	B	C	D	E	F	G	H	I	J	N	O	Q <sub>1</sub>	Q <sub>2</sub>	Q <sub>3</sub>	
Part A <sub>1</sub> Raw Score																
Part A <sub>2</sub> Raw Score																
Form A Raw Score = (A <sub>1</sub> + A <sub>2</sub> )																
Standard Score																
PROFILE																
IN																
STENS																
FACTOR	A	B	C	D	E	F	G	H	I	J	N	O	Q <sub>1</sub>	Q <sub>2</sub>	Q <sub>3</sub>	

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# What You Do and What You Think

Print Your Name: First \_\_\_\_\_ Last \_\_\_\_\_

Your Age \_\_\_\_\_ Grade in School \_\_\_\_\_ Boy or Girl \_\_\_\_\_

Read each statement and mark an  on the side that fits you better. Some questions will not have the words just the way you want them but mark every one the best you can. You may ask for help if you don't know a word. Just raise your hand and the teacher will come to your desk. Do not work long on one question. Mark it and go right on to the next one. MARK EVERY ONE. Most of the questions have two boxes to choose from but other questions have three boxes. Always look at ALL the boxes and pick just one of them for your answer.

1. Do you finish your school work quickly  or  does it take you too long
2. When losing a game, do you sometimes give up and save your energy  or  always play harder
3. Can you easily persuade your friends to accept your plans  or  is it difficult.
4. Do you think many children do better work than you  or  are you as good as anyone else
5. If the teacher lets another child do a job you want to do, do you feel badly  or  soon forget about it.
6. Do grown-ups think you are naughty  or  well-behaved

7. Do you find other children take advantage of you  or  are they kind to you
8. Do you make a lot of mistakes  or  just a few
9. Do people like your ideas  or  do they not like them
10. If you got lost, would you know what to do  or  would you be scared

**GO RIGHT ON TO THE NEXT PAGE.**

DO NOT WRITE BELOW THIS LINE.

FACTOR	A	B	C	D	E	F	G	H	I	J	N	O	Q <sub>1</sub>	Q <sub>2</sub>
Part A1 Raw Score														
Part A2 Raw Score														
Form A Raw Score = (A1 + A2)														
Standard Score														
10														
9														
8														
7														
6														
5														
4														
3														
2														
1														
FACTOR	A	B	C	D	E	F	G	H	I	J	N	O	Q <sub>1</sub>	Q <sub>2</sub>

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## APPENDIX J

## PRIMARY FACTORS ON THE 16PF

FACTOR	Standard Score	LOW SCORE DESCRIPTION	HIGH SCORE DESCRIPTION
A		RESERVED, DETACHED, CRITICAL, COOL (Sizothymia)	OUTGOING, WARMHEARTED, EASY-GOING, PARTICIPATING (Affectothymia or cyclothymia)
B		LESS INTELLIGENT, CONCRETE-THINKING (Lower scholastic mental capacity)	MORE INTELLIGENT, ABSTRACT-THINKING, BRIGHT (Higher scholastic mental capacity)
C		AFFECTED BY FEELINGS, EMOTIONALLY LESS STABLE, EASILY UPSET (Lower ego strength)	EMOTIONALLY STABLE, FACES REALITY, CALM (Higher ego strength)
E		HUMBLE, MILD, OBEYING, CONFORMING (Submissiveness)	ASSERTIVE, INDEPENDENT, AGGRESSIVE, STUBBORN (Dominance)
F		SOBER, PRUDENT, SERIOUS, TACITURN (Desurgency)	HAPPY-GO-LUCKY, HEEDLESS, GAY, ENTHUSIASTIC (Surgency)
G		EXPEDIENT, A LAW TO HIMSELF, BY-PASSES OBLIGATIONS (Weaker superego strength)	CONSCIENTIOUS, PERSEVERING, STAID, RULE-BOUND (Stronger superego strength)
H		SHY, RESTRAINED, DIFFIDENT, TIMID (Threctia)	VENTURESOME, SOCIALLY BOLD, UNINHIBITED, SPONTANEOUS (Parmia)
I		TOUGH-MINDED, SELF-RELIANT, REALISTIC, NO-NONSENSE (Harrtia)	TENDER-MINDED, DEPENDENT, OVER-PROTECTED, SENSITIVE (Premia)
L		TRUSTING, ADAPTABLE, FREE OF JEALOUSY, EASY TO GET ON WITH (Alaxia)	SUSPICIOUS, SELF-OPINIONATED, HARD TO FOOL (Protension)
M		PRACTICAL, CAREFUL, CONVENTIONAL, REGULATED BY EXTERNAL REALITIES, PROPER (Praxernia)	IMAGINATIVE, WRAPPED UP IN INNER URGENCIES, CARELESS OF PRACTICAL MATTERS, BOHEMIAN (Autia)
N		FORTHRIGHT, NATURAL, ARTLESS, SENTIMENTAL (Artlessness)	SHREWD, CALCULATING, WORLDLY, PENETRATING (Shrewdness)
O		PLACID, SELF-ASSURED, CONFIDENT, SERENE (Untroubled adequacy)	APPREHENSIVE, WORRYING, DEPRESSIVE, TROUBLED (Guilt proneness)
Q <sub>1</sub>		CONSERVATIVE, RESPECTING ESTABLISHED IDEAS, TOLERANT OF TRADITIONAL DIFFICULTIES (Conservatism)	EXPERIMENTING, CRITICAL, LIBERAL, ANALYTICAL, FREE-THINKING (Radicalism)
Q <sub>2</sub>		GROUP-DEPENDENT, A "JOINER" AND SOUND FOLLOWER (Group adherence)	SELF-SUFFICIENT, PREFERS OWN DECISIONS, RESOURCEFUL (Self-sufficiency)
Q <sub>3</sub>		CASUAL, CARELESS OF PROTOCOL, UNTIDY, FOLLOWS OWN URGES (Low integration)	CONTROLLED, SOCIALLY-PRECISE, SELF-DISCIPLINED, COMPULSIVE (High self-concept control)
Q <sub>4</sub>		RELAXED, TRANQUIL, TORPID, UNFRUSTRATED (Low ergic tension)	TENSE, DRIVEN, OVERWROUGHT, FRETFUL (High ergic tension)

## APPENDIX J - Continued

## Secondary Factors on the 16PF

Low Score Definition	Factor	High Score Definition
Invia (intraversion)	Q <sub>1</sub> <sup>a</sup>	Exvia (extraversion)
Adjustment	Q <sub>11</sub> <sup>a</sup>	Anxiety
Pathemia	Q <sub>111</sub> <sup>a</sup>	Cotertia (tough poise)
Subduedness	Q <sub>1v</sub> <sup>a</sup>	Independence
Naturalness	Q <sub>v</sub>	Discreteness
Cool Realism	Q <sub>v1</sub>	Prodigal Subjectivity

a. These secondary factors are also present on the HSPQ and the CPQ.



APPENDIX K

Primary Factors of the HSPQ

LOW SCORE DESCRIPTION	STANDARD TEN SCORE (STEN)	HIGH SCORE DESCRIPTION
RESERVED, DETACHED, CRITICAL, ALOOF, STIFF (Sizothymia)	1 2 3 4 5 6 7 8 9 10	WARMHEARTED, OUTGOING, EASY-GOING, PARTICIPATING (Affectothymia, formerly cyclothymia)
DULL, CONCRETE-THINKING (Lower intelligence)	A	BRIGHT, ABSTRACT-THINKING (Higher intelligence)
AFFECTED BY FEELINGS, EMOTIONALLY LESS STABLE, EASILY UPSET, CHANGEABLE (Lower ego strength)	B	EMOTIONALLY STABLE, MATURE, FACES REALITY, CALM (Higher ego strength)
UNDEMONSTRATIVE, DELIBERATE, INACTIVE, SLODDY (Phlegmatic temperament)	C	EXCITABLE, IMPATIENT, DEMANDING, OVERACTIVE, UNRESTRAINED (Excitability)
OBEDIENT, MILD, EASILY LED, DOCILE, ACCOMMODATING (Submissiveness)	D	ASSERTIVE, AGGRESSIVE, COMPETITIVE, STUBBORN (Dominance)
SOBER, TACITURN, SERIOUS (Desurgency)	E	ENTHUSIASTIC, HEEDLESS, HAPPY-EGO-LUCKY (Surgency)
DISREGARDS RULES, EXPEDIENT (Weaker superego strength)	F	CONSCIENTIOUS, PERSISTENT, MORALISTIC, STAID (Stronger superego strength)
SHY, TIMID, THREAT-SENSITIVE (Threctia)	G	ADVENTUROUS, "THICK-SKINNED," SOCIALLY BOLD (Fornia)
TOUGH-MINDED, REJECTS ILLUSIONS (Harrta)	H	TENDER-MINDED, SENSITIVE, CLINGING, OVER-PROTECTED (Premtia)
ZESTFUL, LIKES GROUP ACTION (Zeppia)	I	CIRCUMSPECT INDIVIDUALISM, REFLECTIVE, INTERNALLY RESTRAINED (Coasthentia)
SELF-ASSURED, COMPLACENT, SECURE, PLACID, SERENE (Untroubled adequacy)	J	APPREHENSIVE, SELF-REPROACHING, INSECURE, WORRYING, TROUBLED (Guilt proneness)
SOCIABLY GROUP-DEPENDENT, A "JOINER", AND SOUND FOLLOWER (Group dependency)	K	SELF-SUFFICIENT, PREFERENCES OWN DECISIONS, RESOURCEFUL (Self-sufficiency)
UNCONTROLLED, LAX, FOLLOWS OWN URGES, CARELESS OF SOCIAL RULES (Low self-sentiment integration)	L	CONTROLLED EXACTING WILL POWER, SOCIALLY PRECISE, COMPULSIVE, (High strength of self-sentiment)
RELAXED, TRANQUIL, TORPID, UNFRUSTRATED, COMPOSED (Low-ergic tension)	M	TENSE, FRUSTRATED, DRIVEN, OVERWROUGHT, FRETFUL (High-ergic tension)

APPENDIX L

Primary Factors of the CPQ

LOW SCORE DESCRIPTION	STANDARD TEN SCORE (STEN)										HIGH SCORE DESCRIPTION
	1	2	3	4	5	6	7	8	9	10	
<b>RESERVED, DETACHED, CRITICAL, COOL</b> (Sizothymia)	•	•	•	•	•	•	•	•	•	•	<b>OUTGOING, WARMHEARTED, EASY-GOING, PARTICIPATING</b> (Affectothymia, formerly cyclothymia)
<b>LESS INTELLIGENT, CONCRETE-THINKING</b> (Lower scholastic mental capacity)	•	•	•	•	•	•	•	•	•	•	<b>MORE INTELLIGENT, ABSTRACT-THINKING, BRIGHT</b> (Higher scholastic mental capacity)
<b>AFFECTED BY FEELINGS, EMOTIONALLY LESS STABLE, EASILY UPSET</b> (Lower ego strength)	•	•	•	•	•	•	•	•	•	•	<b>EMOTIONALLY STABLE, FACES REALITY; CALM, MATURE</b> (Higher ego strength)
<b>PHLEGMATIC, DELIBERATE, INACTIVE, STODGY</b> (Phlegmatic temperament)	•	•	•	•	•	•	•	•	•	•	<b>EXCITABLE, IMPATIENT, DEMANDING, OVERACTIVE</b> (Excitability)
<b>OBEDIENT, MILD, ACCOMMODATING, CONFORMING</b> (Submissiveness)	•	•	•	•	•	•	•	•	•	•	<b>ASSERTIVE, INDEPENDENT, AGGRESSIVE, STUBBORN</b> (Dominance)
<b>SOBER, PRUDENT, SERIOUS, TACITURN</b> (Desurgency)	•	•	•	•	•	•	•	•	•	•	<b>HAPPY-GO-LUCKY, IMPULSIVELY LIVELY, GAY, ENTHUSIASTIC</b> (urgency)
<b>EXPEDIENT, EVADES RULES, FEELS FEW OBLIGATIONS</b> (Weaker superego strength)	•	•	•	•	•	•	•	•	•	•	<b>CONSCIENTIOUS, PERSEVERING, STAIID, RULE-BOUND</b> (Stronger superego strength)
<b>SHY, RESTRAINED, DIFFIDENT, TIMID</b> (Threctia)	•	•	•	•	•	•	•	•	•	•	<b>VENTURESOME, SOCIALLY BOLD, UNINHIBITED, SPONTANEOUS</b> (Parric)
<b>TOUGH-MINDED, SELF-RELIANT, REALISTIC, NO-NONSENSE</b> (Harric)	•	•	•	•	•	•	•	•	•	•	<b>TENDER-MINDED, DEPENDENT, OVER-PROTECTED, SENSITIVE</b> (Premial)
<b>VIGOROUS, GOES READILY WITH GROUP, ZESTFUL, GIVEN TO ACTION</b> (Zeppia)	•	•	•	•	•	•	•	•	•	•	<b>CIRCUMSPECT, UNWILLING TO ACT WITH GROUP, REFLECTIVE, GUARDED</b> (Coasthenia)
<b>FORTHRIGHT, NATURAL, ARTLESS, SENTIMENTAL</b> (Artlessness)	•	•	•	•	•	•	•	•	•	•	<b>SHREWD, CALCULATING, ASTUTE, CANNY, WARY</b> (Shrewdness)
<b>PLACID, CONFIDENT, SERENE</b> (Untroubled adequacy)	•	•	•	•	•	•	•	•	•	•	<b>APPREHENSIVE, WORRYING, DEPRESSED, TROUBLED</b> (Guilt proneness)
<b>UNDISCIPLINED SELF-CONFLICT, FOLLOWS OWN URGES, CARELESS OF PROTOCOL</b> (Low integration)	•	•	•	•	•	•	•	•	•	•	<b>CONTROLLED, SOCIALLY PRECISE, FOLLOWING SELF IMAGE</b> (High self-concept control)
<b>RELAXED, TRANQUIL, TORPID, UNFRUSTRATED</b> (Low erotic tension)	•	•	•	•	•	•	•	•	•	•	<b>TENSE, FRUSTRATED, DRIVEN, OVERWROUGHT</b> (High erotic tension)

## APPENDIX M

## STRUCTURED INTERVIEW - ADULT VERSION

## ORALLY ADMINISTERED

"I would like to obtain some information about you and the things that are happening to you, this is completely confidential".

"I am going to tape record our conversation so that I won't have to copy what you say and to save time."

Number-

Cystinuric-

Sex-

Non-cystinuric-

Birthdate

Marital status-

Occupation-

Highest grade completed-

Height-

Weight-

1. How is your health? Eyesight? Hearing? Memory?
2. How is your appetite?
3. Have you ever undergone a major operation?
4. Could you describe your present living arrangements, e.g., do you live at home, own your own home, rent an apartment?
5. Describe your own family, your parents, brothers and sisters.

Tell me about them.

Do you still do things with them?

What?

How do they treat you?

6. Could you give me a brief account of your education?

Do you or did you like school?

What did you like and dislike?

Did you have any problems at school?

e.g., some people have difficulty learning to read, to write,  
to spell properly, or to do arithmetic.

How did you get along with your  
teachers, with your classmates?

Did you participate in extra-  
curricular activities in school? What?

7. What do you do now when you are not working (studying)?

Any hobbies or recreations?

Do you have close friends?

Are most of your friends men or women,  
do you prefer the company of men or women?  
Why?

What are your relations like with the  
opposite sex?

Do you or did you ever like to do things that are dangerous?

8. Do you (did you) belong to any social groups? What are (were)  
your functions?

9. Describe the jobs you have had.

10. What kind of plans do you have for the future?

11. Do you ever feel or did you ever feel ill at ease or bothered  
or nervous when you are in a crowd? a bus? a train? an elevator?

a tunnel? a closed room?

12. Is there any other place or situation that makes you feel uncomfortable? Do you try to keep away from it(them)?

13. Would you consider yourself to be a nervous person?

14. How do your co-workers treat you?

neighbours?

classmates?

15. In general, how do you feel about the way things are going for you?

16. How is your mood these days?

17. If you had a friend who was close to you and knew you very well how would he or she describe you as a person; what would they say about you?

## APPENDIX N

## STRUCTURED INTERVIEW-CHILDREN'S VERSION

## ORALLY ADMINISTERED

"I would like to get some information about you and the things that are happening to you, I am going to tape record what we say so that we can save time".

Number-

Cystinuric-

Sex-

Non-cystinuric-

Birthdate-

Marital status-

Occupation-

Highest grade completed-

Height-

Weight-

1. How is your health, how are you feeling?

2. How is your appetite, or do you like to eat?

3. Did you ever have an operation?

4. How many people live in your house?

5. Tell me about your family? How do they treat you?

Do you have any brothers or sisters who are not living at home?

Tell me about them.

6. Tell me about school.

Do you like it?

What do you like? Not like?

What problems do you or did you have in



school?

e.g., some people find it hard learning to read, to write, to spell properly or to do arithmetic. How about you?

Tell me about your teachers.

What about your classmates?

7. What do you do when you are not at school?

Any hobbies?

Tell me about your friends, do you

have many friends?

Are most of your friends boys or girls?

How do you feel about girls(boys)?

Do you or did you ever like to do things that are dangerous?

8. Did you ever belong to any groups? What do you (or did you) do in the group?

9. Did you ever have any jobs? Tell me about them.

10. What would you like to do when you get older?

11. Do you (did you) ever feel bothered or nervous when you're in a crowd, a bus, a train, an elevator, a tunnel, or in a closed room?

12. Is there anything or anything else that makes you feel uncomfortable? What do you do about it? Do you stay away?

13. Would you say that you're a nervous person?

14. How do you feel about the way things are going for you?

15. How is your mood?

16. If you had a friend who knew you very well how would he or she describe you, what would they say about you?

APPENDIX O  
DESIRABILITY SCALE

The total score is indicative of the desirability of an individual's qualities, activities, and situation as presented by him(her) in the interview.

The greater the negative score the less the desirability.

Question List Item	Value
1 . 1. indicates good health	0
2. fairly good health	0
3. feels tired sleepy or without energy	-1
4. has various aches or pains or physical dysfunctions.	-1
5. expresses dissatisfaction with his size or strength	-1
6. says his body is decaying	-1
7. dissatisfied with the appearance of body or part of his body	-1
8. eyes satisfactory	0
9. eyes not satisfactory	-1
10. hearing satisfactory	0
11. hearing not satisfactory	-1
12. memory O.K.	0
13. mentions memory impaired or that he keeps forgetting things	-1

Question	List Item	Value
	14. reports difficulty recalling important details of past experience	-1
	15. tells of fit, seizure or lack of consciousness	-1
2	16. indicates good appetite	0
	17. indicates poor appetite	-1
3.	18. has had major operation	-1
	19. has not had major operation	0
4.	20. no scoring	0
5.	21. enjoys family and likes members	0
	22. expresses negative attitudes towards family members	-1
	23. says he is unwilling to see any member of his immediate family	-1
	24. expresses jealousy, rivalry, or bitter envy toward family member	-1
	25. says does not care what happens to his family	-1
	26. tells how his family has mistreated or harmed him.	-1
	27. expresses hatred for relative	-1
6.	28. high school complete and above	0
	29. student now	0
	30. not a student, not finished high school	-1

Question	List Item	Value
	31. positive attitude towards school	0
	32. valid criticism of school	0
	33. highly critical of school	-1
	34. minor nonspecific difficulties at school	0
	35. a distinct behavior problem at school	-1
	36. any specific or not so specific learning difficulty in learning to read	-1
	37. write	-1
	38. spell	-1
	39. do arithmetic	-1
	40. other	-1 each
	41. good relationship with teachers	0
	42. poor relationship with teachers	-1
	43. good relationship with classmates	0
	44. poor relationship with classmates	-1
	45. participated in extra-curricular activities	0
	46. did not participate in extra-curricular activities	-1
7.	47. mentions socially acceptable activities	0
	48. socially unacceptable activities	-1
	49. mentions that nothing interests him	-1
	50. expresses feeling of inadequacy	-1

Question	List Item	Value
	51. has close <sup>d</sup> friends	0
	52. no close friends	-1
	53. constructive interest in opposite sex	0
	54. uninterested in opposite sex	-1
	55. becomes flustered when asked this and expresses feeling of fear or discomfort	-1
	56. says has difficulty getting along	-1
	57. <sup>b</sup> reports deficiency in sexual performance	-1
	58. other sexual problem	-1
	59. expresses feeling of guilt	-1
	60. enjoys doing dangerous things	-1
	61. indicates is thinking of or tried to kill or harm self	-1
	62. does not do dangerous things	0
8.	63. belongs, socially acceptable group	0
	64. does not belong and is not interested	0
	65. does not belong to groups but is dissatisfied	-1
9.	66. if applicable, has held a job for more than one year	0
	67. is a job hopper	-1
10.	68. constructive plans for future	0
	69. no plans for future	-1
	70. expresses a negative attitude towards future	-1

Question	List Item	Value	
11.	71. feels ill at ease in a crowd	-1	
	72. bus	-1	
	73. train	-1	
	74. elevator	-1	
	75. tunnel	-1	
	76. closed room	-1	
	77. other	-1 each	
12.	78. keeps away from place or situation which is uncomfortable	-1	
	79. does not stay away, copes with the anxiety	0	
13.	80. says he is a nervous person	-1	
	81. says he is not a nervous person	0	
14.	82. gets along well with neighbours, coworkers	0	
	83. reports getting angry when criticized	-1	
	84. tells of being mistreated	-1	
	85. tells that someone talks about or ridicules him	-1	
	86. says he believes someone wishes to harm him	-1	
	15.	87. satisfied with the way things are going	0
		88. dissatisfied with the way things are going	-1
89. intense regret expressed for something done or not done		-1	

## APPENDIX 0 - Continued

99

Question	List Item	Value
	90. talks of serious personal problem in flat unemotional manner	-1
16.	91. mood satisfactory	0
	92. feels elated or high or depressed	-1
	93. feels is getting nowhere	-1
17.	94. describes self with positive characteristics	+1 each
	95. describes self with negative characteristics	-1 each

APPENDIX P

DATA AND DATA ANALYSES



TABLE P-1

Summary of Mean I.Q. Scores and Standard  
Deviations of Cystinurics and Controls

Intelligence Scale	Group			
	Cystinuric		Control	
	Mean	SD	Mean	SD
Verbal	96.58	14.98	91.5	11.91
Performance	97.17	15.95	97.1	9.13
Full	96.42	15.97	93.67	10.27

TABLE P-2

Experimental Design and Standard Scores used in Analysis of  
Variance on Intelligence Scale Subtests

Pair Number	Cystinurics										Controls										
	Subtests 1-10 <sup>a</sup>										Subtests 1-10 <sup>a</sup>										
1	9	6	8	12	10	9	12	12	11	13	8	7	8	10	10	8	14	16	8	8	
2	18	13	14	12	10	14	17	13	12	10	14	13	9	14	13	11	8	11	11	8	
3	11	11	14	13	12	10	12	10	10	10	9	10	10	10	8	10	9	11	5	10	
4	13	11	8	11	8	12	12	13	9	11	9	9	4	11	7	11	6	7	11	12	
5	12	10	11	8	10	8	9	13	11	13	10	11	10	10	9	9	8	8	9	7	
6	7	8	7	7	6	5	9	7	4	5	5	4	5	5	5	3	8	8	7	9	
7	6	5	8	9	13	7	8	6	7	3	6	5	10	10	7	5	11	6	9	7	
8	4	2	5	7	5	7	7	6	7	3	6	6	9	5	7	8	10	8	10	8	
9	8	13	13	8	4	10	10	9	10	7	12	9	9	12	9	11	9	12	10	11	
10	11	10	10	10	8	7	9	9	7	7	10	11	8	8	4	9	13	11	13	10	
11	12	12	14	15	8	11	13	13	11	7	10	11	11	11	9	6	13	13	7	10	9
12	6	10	5	12	7	7	13	11	10	9	6	9	9	12	9	8	6	12	9	11	15

a. Subtests 1-10 include Information, Comprehension, Arithmetic, Similarities, Digit Span Vocabulary, Picture Completion, Block Design, Picture Arrangement, and Object Assembly respectively.

TABLE P-3  
 Experimental Design and Standard Scores used in Analysis of  
 Variance on Personality Source Factors

Pair Number	Cystinurics													Controls									
	Personality Source Factors													Personality Source Factors									
	A	B	C	E	F	G	H	I	O	Q <sub>3</sub>	Q <sub>4</sub>	A	B	C	E	F	G	H	I	O	Q <sub>3</sub>	Q <sub>4</sub>	
1	5	6	7	6	2	5	5	6	5	6	5	4	7	6	6	4	7	4	8	3	5	5	
2	5	8	5	8	7	3	8	7	4	6	5	7	8	7	3	4	7	8	6	3	10	4	
3	5	4	3	7	5	6	6	5	5	6	4	4	5	3	4	7	4	2	6	9	8	7	
4	5	6	2	7	6	5	5	3	8	5	6	3	4	6	8	6	10	6	6	6	4	7	
5	3	7	5	5	5	5	3	4	6	2	9	5	2	4	2	2	9	1	7	3	7	5	
6	7	3	3	3	5	7	5	6	8	1	5	5	2	6	4	3	6	4	9	8	7	8	
7	4	3	6	5	4	6	5	5	7	7	7	3	4	4	4	7	2	7	4	6	6	4	
8	4	2	6	5	4	3	5	8	8	9	6	5	2	9	3	5	4	8	5	1	10	5	
9	4	2	7	4	6	6	6	6	5	7	4	9	6	8	3	7	6	3	6	5	6	6	
10	4	4	2	1	3	6	3	3	4	7	4	8	6	4	4	5	6	8	5	5	5	7	
11	1	8	2	7	9	3	3	5	8	2	9	2	3	7	5	6	2	3	8	9	1	9	
12	5	3	9	5	4	4	8	4	4	5	3	10	3	6	1	1	8	3	8	6	8	4	

TABLE P-4

Mean Standard Scores and Standard Deviations of  
Cystinurics and Controls on each Primary and  
Secondary Personality Factor

Personality Factor Code	Group			
	Cystinuric		Control	
	Mean	SD	Mean	SD
A	4.33	1.43	5.4	2.53
B	4.66	2.22	4.3	2.05
C	4.75	2.33	5.86	1.80
E	5.25	1.95	3.9	1.83
F	5.0	1.85	4.67	2.0
G	4.9	1.37	5.9	2.53
H	5.17	1.69	4.67	2.0
I	5.17	1.52	6.5	1.50
O	6.0	1.7	5.3	2.53
Q <sub>3</sub>	5.25	2.41	6.4	2.53
Q <sub>4</sub>	5.6	1.92	5.9	1.67
Q <sub>1</sub>	4.34	1.98	4.65	1.08
Q <sub>11</sub>	5.56	1.52	5.53	1.19
Q <sub>111</sub>	5.66	1.02	4.56	1.45
Q <sub>1V</sub>	5.23	1.67	4.86	1.79

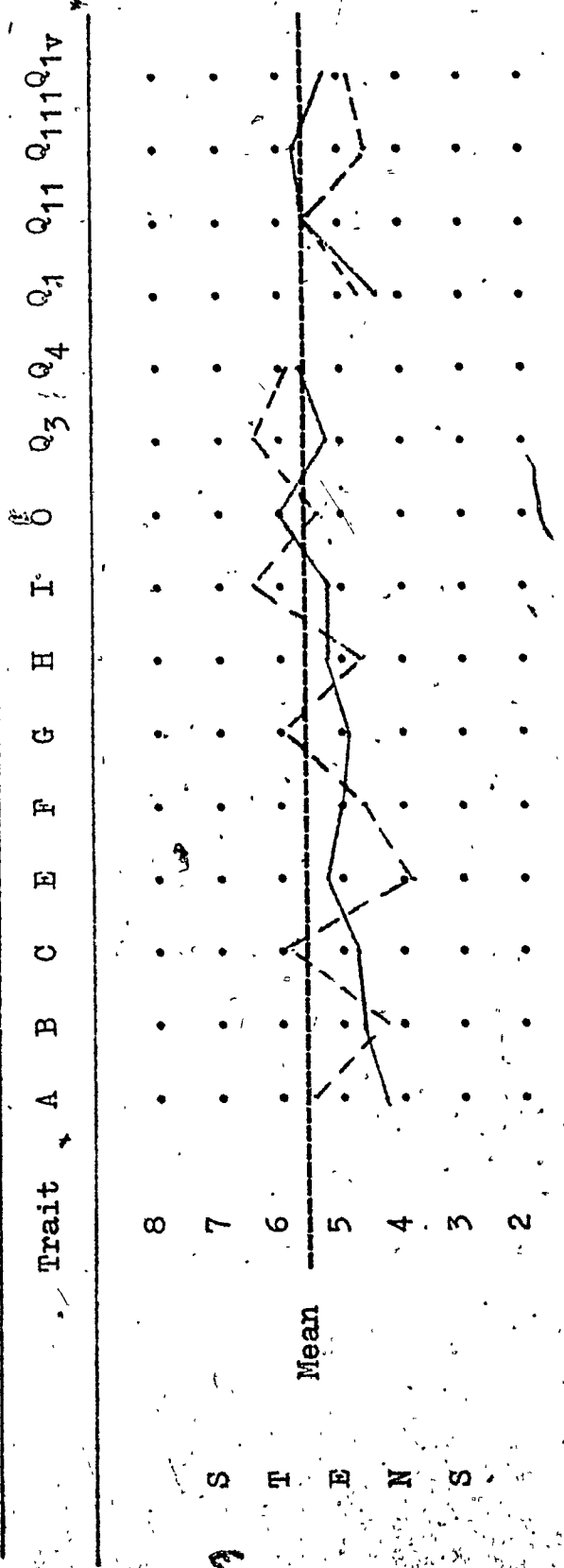


Fig. P-1. Comparison of personality profiles on primary and secondary traits for Cystinurics (solid line) and controls (broken line).

TABLE P-5

Experimental Design and Standard Scores used in Analysis of  
Variance on Secondary Personality Factors

Pair Number	Cystinurics				Controls			
	Secondary Personality Factors				Secondary Personality Factors			
	Q <sub>1</sub>	Q <sub>11</sub>	Q <sub>111</sub>	Q <sub>1V</sub>	Q <sub>1</sub>	Q <sub>11</sub>	Q <sub>111</sub>	Q <sub>1V</sub>
1	3.5	4.5	5.2	6.2	4.2	5.7	4.5	4.7
2	6.2	4.4	6.8	7.1	5.8	3.2	4.8	4.3
3	5.7	4.9	6.0	5.6	3.4	7.3	4.3	5.1
4	5.2	7.3	7.0	3.1	5.3	5.7	6.6	8.4
5	4.8	6.9	6.1	6.0	3.3	5.1	3.8	3.7
6	2.5	7.4	3.5	3.2	2.7	5.9	3.5	6.9
7	3.6	5.7	5.8	6.3	5.4	6.0	6.5	4.1
8	3.0	3.3	4.8	6.9	5.3	3.9	5.6	6.4
9	4.9	3.8	4.6	5.6	5.6	5.4	1.7	1.6
10	1.6	5.3	5.3	2.3	5.8	6.2	4.8	3.3
11	5.4	7.8	6.6	6.7	4.9	7.2	5.7	5.6
12	5.7	4.4	6.2	3.8	4.1	4.8	2.9	4.2

TABLE P-6  
Summary of Inter-rater  
Reliability for the  
Desirability Scale

Rater Comparisons	$r_s$
1 <sup>a</sup> vs 2 <sup>b</sup>	.794**
1 vs 3 <sup>c</sup>	.691**
2 vs 3	.646**

a, b Independent naive raters

c The interviewer

\*\* p < .01

TABLE P-7

Summary of Analyses on each Factor Comparing Mean Standard Scores of the Total Cystinuric Sample with the Constant Normative Mean of 5.5 and SD of two

Personality Factor Code	Mean	SD	Z <sup>a</sup>
A	4.92	1.81	1.48
B	5.5	2.04	0
C	4.81	2.30	1.76
E	5.62	1.87	.31
F	5.42	1.88	.20
G	4.65	1.95	2.17*
H	5.04	2.10	1.12
I	5.92	1.95	1.07
O	5.88	2.46	.97
Q <sub>3</sub>	5.08	2.31	1.07
Q <sub>4</sub>	5.73	2.32	.59

a Two-tail test

\*  $p < .05$



Source Trait A B C E F G H I O Q<sub>3</sub> Q<sub>4</sub>

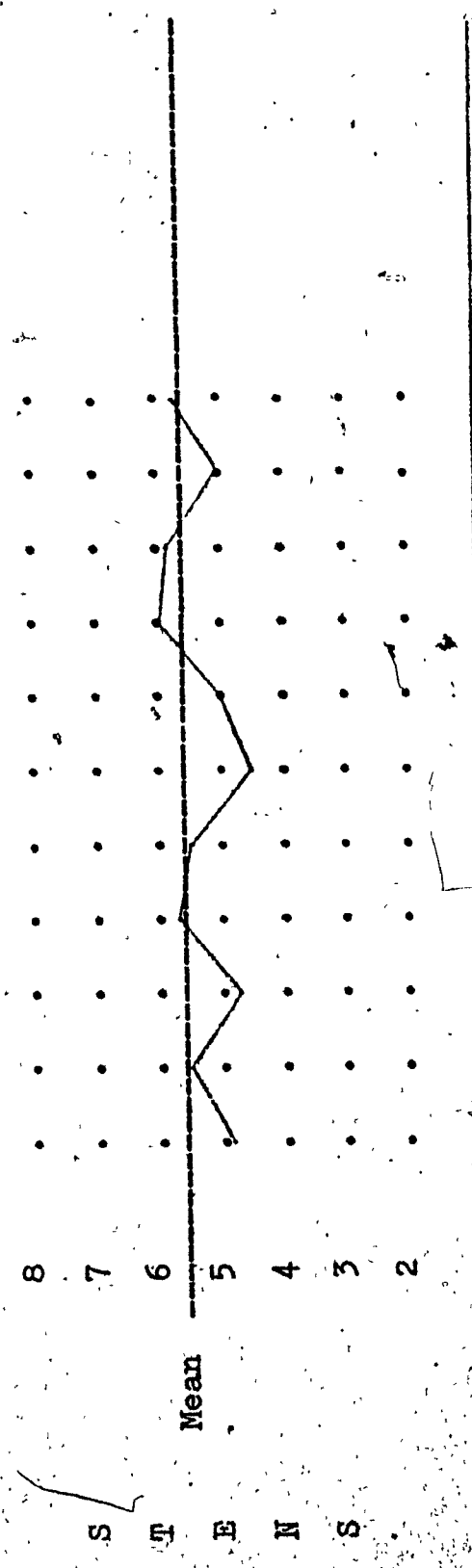


Fig. P-2. Personality profile of mean scores on primary traits for 26 Cystinurics.