AN ABSTRACT OF THE THESIS OF

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The purpose of this study was to determine the possible problem areas in families with a diet-treated phenylketonuric child. Phenylketonuria is an inborn error of metabolism, which in the past caused mental retardation as well as behavioral disturbances. Initially the discovery of diet therapy eliminated the behavioral disturbances, but not the mental retardation in phenylketonurics, primarily because of the timing of the diagnosis. In recent years, however, a test has been developed which identifies the disease in early infancy, and with early use of the diet therapy, both the mental retardation and the behavioral symptoms have been eliminated.

However, because of the strictness of the diet, and as a result of studies which indicate that the diet-treated phenylketonuric child may be slower in growth and lower in I. Q. than children who do not have phenylketonuria, there exists the possibility that there may be problems within the family related to the presence of the

phenylketonuric child.

In the review of the literature, areas of "normal" childhood, which could be problem areas, were discussed, such as: learning, parent-child interactions, dependency and overprotection, language development, peer relationships, family relationships, and family developmental tasks. These areas were then discussed in terms of the phenylketonuric child and his family.

It would be reasonable to assume that the presence of the phenylketonuric child in the family would create a discrepancy with regard to the concept of the child formed by the parents before the birth of the child; create parental anxiety because of the nature of the illness, and affect family food patterns because of the strictness of the diet. It would be reasonable to assume that since children with a chronic illness are often dependent, and their parents often show signs of overprotection, that this would be true in the case of the phenylketonuric child, also. Because of the possibility of reduced contact with peers, and the possible lower I. Q. of the phenylketonuric child, it would be reasonable to assume that this child might be slower in language development. Because of the strictness of the diet, and the reported slower growth of the phenylketonuric child, the child might be smaller than his "normal" age-mates, show signs of fatigue more quickly and therefore, be adversely affected in his social development. Because of the possible presence of fear-producing stimuli in the child's

environment, such as the fear and anxiety of the parents, the child might show signs of fear and anxiety. Children with chronic illness generally show signs of emotional instability, and the phenylketonuric child could be expected to follow this pattern. Any of these complications could affect the child's self-concept and self-acceptance adversely, and could also be expected to affect his peer relationships adversely.

The phenylketonuric child could be expected to affect family relationships in that parental concern could be directed toward this child with less attention given to other children in the family. Also, social relationships outside the family could be affected. The phenylketonuric child could be expected to affect the family developmental tasks in that the added task of adjusting to the child and his problems could take priority over other tasks that the family would be expected to complete.

For the purpose of exploring these considerations to determine whether problems in these areas do exist, an interview schedule was formulated for use in interviewing families with a phenylketonuric child five years of age or younger. Some of the questions for the interview schedule were taken from the literature dealing directly with the known problems of the phenylketonuric child. Others were taken from the normative-descriptive literature involving problem areas in "normal" childhood, as discussed above, and others were questions

which were inferred from the literature concerning the family.

On the assumption that the expected problems would exist, a discussion of possible solutions to the problems was given.

Problems of Families With a Diet-treated Phenylketonuric Child

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BACKGROUND OF STUDY

The inborn error of metabolism, phenylketonuria, was first discovered by Folling in 1934 (Wooley, 1965). It was not until much later that it was found that the effects of phenylketonuria could be controlled through diet. Later, various methods of early detection of phenylketonuria were discovered. Phenylketonuria is said to occur once in every 10, 000 births (Painter, 1965).

The first test used was the addition of ferric chloride to a urine specimen taken from the infant, or a drop of ferric chloride placed on the wet diaper of the infant. However, this test proved unsuccessful in that it was not reliable until the infant was four to six weeks in age, and as shown from other studies, brain damage could have occurred by this time (Kuegel, 1966).

Later, a bacterial inhibition assay test was developed by Guthrie, which used a drop of blood taken from the infant during the first three days of life. Shortly after this, LaDu developed an enzymatic spectrophotometric test, which proved to be as accurate as the Guthrie test, but required a great amount of time for each individual test, and required expensive equipment. Tests on the effectiveness of both the LaDu and Guthrie tests were done in Iowa and Rhode

Island on 2, 607 infants, and although both tests proved to be accurate, the Guthrie test was most applicable to mass screening because of its efficiency (Kuegel, 1966). The Guthrie test is the one used in Oregon, at present (Painter, 1965).

Testing for phenylketonuria began in Oregon in 1962. However, it was not until 1965 that the Oregon State Board of Health began a mass testing program for the detection of phenylketonuria, and several other inborn errors of metabolism. At that time, there were two other states with this type of program, namely Massachusetts and California (Ostergren, 1964). Presently, there are 39 other states using the screening program (Floyd, 1967).

Review of Metabolism

In order to understand the immensity of the problems faced by the child with phenylketonuria and his family, a review of metabolism might be worthwhile. All of the food we eat, with the exception of some things like coffee and tea, is broken down chemically in the body, both in the digestive tract and in the various organs and cells of the body. After breaking down these foodstuffs, the body then utilizes them along with oxygen from the air, to provide energy for growth, maintenance and repair of tissues, and regulation of the body processes, including metabolism itself. The body processes occur in a minutely detailed pattern that is so perfectly balanced that if one

pattern is changed in any way, the result can be death for that individual.

Phenylketonuria vs. Metabolism

For a graphic description of the following, the reader may wish to refer to Appendix A.

Phenylketonuria is a result of a defect in the metabolism of the amino acid, phenylalanine. The amino acids are the primary units of proteins. There are twenty amino acids. Eight of them are termed, "essential", meaning that the body cannot manufacture them from other substances. Phenylalanine is one of these eight essential amino acids.

Phenylalanine occurs in 4 to 6% of the naturally occurring proteins (Lyman, 1963). Normally, part of the amount taken into the body is converted into another amino acid, tyrosine. Every reaction in the body is helped in some way by an enzyme, which is a chemical compound in the body, acting as a catalyst in body processes. In phenylketonuria, the enzyme system which normally converts phenylalanine into tyrosine is missing. Tyrosine is not an essential amino acid. The body can either make it from phenylalanine, or obtain it from food. However, since the body is built to convert a specific amount of phenylalanine into tyrosine, when it cannot, the excess phenylalanine is converted into phenylpyruvic acid, phenylacetic acid,

and phenyllactic acid. These products then build up in the blood stream, and in effect, unbalance the system.

In persons with phenylketonuria, large amounts of these three products are found in the urine and blood stream (Hsia, 1959). It has been found that phenylalanine and its by-products in the blood stream inhibit the formation of serotonin. This is a hormone found in the brain, and its exact function is not yet known (Lyman, 1963). It is known that serotonin is made from another amino acid, tryptophan, and the products formed from excess phenylalanine inhibit the enzyme system which forms serotonin.

Symptoms of Untreated Phenylketonuria

I.Q. Range

In the population recorded by Stanbury (1960), approximately 2% had an I.Q. above 60. None of these could be described as normal. Hsia (1959) reports that the majority have an I.Q. of 30 or less and that a high I.Q. is found in less than 1% of all patients. In a study reported by Jervis, 94% had an I.Q. of less than 50 (Hsia, 1959).

Development and Behavioral Symptoms

Because of the fact that it is often difficult to maintain sufficient nourishment in severely retarded people, children with phenylketonuria were often found to be under average weight and height (Stanbury,

1960; Paine, 1957). Some of the terms used to describe the walk of a phenylketonuric person are a "stiff" or "stumbling gait," "short steps" and a "stooping walk" (Stanbury, 1960, p. 332).

> They are restless, fearful, jerky individuals. Their behavior ranges from that of shy, anxious, restless high-grade individuals to the destructive and noisy psychotic episodes observed in 10% (Stanbury, 1960, p. 335).

High-grade patients frequently have night-terrors. The patients have abnormally brisk tendon reflexes and appear always ready to jump. Most phenylketonurics were institutionalized be cause of uncontrollable temper tantrums, hyperactivity, and irritability (Stanbury, 1960).

Many abnormal body movements are seen, such as "pill-rolling movements," "irregular tic-like motions," and "aimless to and fro movements of the fingers." Also frequently seen is the habitual "fiddling of the fingers close before the eyes" (Paine, 1957, p. 296). This may be accompanied by a tailor-sitting position, with a "rhythmic rocking back and forth which may continue for hours" (Stanbury, 1960, p. 335; Paine, 1957, p. 299). Often, the patients sit grinding their teeth, along with these other movements (U. S. Children's Bureau 1964b). Stanbury (1960) says that each patient has a limited repertoire of these movements. If one motion is suppressed, others will take its place. The Babinski and Hoffman reflexes are rare in the infant phenylketonuric (Paine, 1957).

Usual developmental progress like sitting, and walking, may be delayed. Teeth may not appear until after the eleventh month

(Stanbury, 1960). Stanbury goes on to say that at the latest ages at which walking and talking could be expected in a normal child, 35% of the phenylketonurics could not walk, and 63% could not talk, and those who could not walk had no sphincter control. In Paine's (1957) study of 106 patients, he found that the mean age of sitting was between 12 and 15 months; of walking, around 36 months; and of talking, between three and four years.

It was noted that some patients lost skills such as talking as they grew older, which they may have acquired in childhood. The low-grade children produce a "parrot-like speech" which Paine (1957) says they develop as a game, but since this game cannot be converted into words with meaning, the game becomes boring, and consequently, the speech is lost.

Phenylketonuric children had been known to have epileptic seizures (Stanbury, 1960; Knox, 1960; Paine, 1957). The frequency was 26% in the cases reported by the two authors cited in Stanbury's study, 33% of the low-grade individuals in the study by Paine, and 33% in the study by Knox. All seizures were said to subside before adulthood. Stanbury (1960) reports that petit mal seizures after six months of age are frequently recorded. He also noted that prolonged, severe epilepsy is rare, but it has been noted as a complication of the low-phenylalanine diet therapy.

It might be wise to add at this point that epilepsy is not a disease,

but a symptom of an abnormal electrochemical occurrence in the brain. The seizures may affect any individual, and may have their onset at any age. It can be caused by any abnormality in the brain cells, or the body, which causes a chemical imbalance (U. S. Children's Bureau, 1964a). It would then seem reasonable for the phenylketonuric child to have epilepsy, due to the vast amount of chemical imbalance in the system.

Another symptom of the phenylketonuric which has been noted is a deficiency in pigmentation, giving blond hair and blue eyes (Stanbury, 1960). This stands to reason, since the amino acid tyrosine is eventually converted to the pigment, melanin, which is responsible for the coloring in any individual. If the tyrosine metabolism suffers a shortage due to lack of phenylalanine conversion, one would expect some decrease in melanin (Paine, 1957). Again, a graphic description may be found in Appendix A.

Dietary Treatment

According to Lyman (1963), it is difficult to form a phenylalanine free diet, because as already stated, phenylalanine makes up 4 to 6% of naturally occurring protein. It is most common to use a synthetic mixture of amino acids, restricting the phenylalanine. "Lofenalac" and "Ketonil" are the products commonly used. Along with these synthetic products, fruits and vegetables containing 1%

protein, gluten-free bread, sugar, butter, vitamins and minerals are used (Hsia, 1959).

It is continually necessary to assess the amount of phenylalanine needed for each patient. Too much phenylalanine in the blood causes mental retardation, but too little will cause death. According to Lyman (1963), the phenylalanine should be restricted until the blood level returns to normal. Then, supplementary foods may be added to the diet, as long as the blood level remains normal. There will also be variations in the amount of phenylalanine allowed for each child due to the activity of the child. "Adequate control depends upon meeting, but not exceeding phenylalanine requirements and meeting caloric and other nutritional requirements" (Stanbury, 1960, p. 371). Illness can cause the level of phenylalanine in the blood to increase, and the level is then depleted after illness, causing problems in dietary balance. The phenylalanine must also be increased as the growth of the individual demands (Sutherland, 1966). There is also a difficulty encountered with lowered phenylalanine in the body, as the body will break down its own protein to get phenylalanine if the dietary supply is depleted (Stanbury, 1960).

Effect of Dietary Treatment on Phenylketonuric Patients

The dietary treatment of patients with phenylketonuria clears up all of the previously mentioned symptoms except the mental

retardation (Stanbury, 1960). If dietary treatment is begun from three days to eleven weeks, the development of the child is considered normal (Sutherland, 1966). If treatment begins after this, the results are less favorable. Lyman (1963) says that treatment begun after six months is less favorable. Brain damage cannot be prevented, but some improvement in I.Q. is shown. After five years of age, even this increase is negligible, but progressive mental decline is avoided. Stanbury (1960) reports severe epileptic seizures at the onset of the diet in already-damaged patients. The U. S. Children's Bureau (1964b) reports seizures when the diet therapy is discontinued.

There are varied opinions as to when the diet should or could be discontinued. Knox (1960) suggests that 24 months is the critical period for irreversible brain damage, and that after this, the patients need not be kept on the diet. Horner et al., (1959) stopped the diets at two months, 18 months, and 30 months, and found no deterioration, but he also suggested that perhaps these individuals would have achieved their mental level without the aid of the diet. Lyman (1963) suggests continuing the diet for at least three or four years, and perhaps up to age eight or nine. Bickel suggests continuing the diet until adolescence (U. S. Children's Bureau, 1964b).

D. W. Woolley (1965), who was with the Rockefeller Institute until his recent death, has suggested that discontinuation of the diet may be dangerous at any age. His reasoning comes from research

involving serotonin, which was mentioned earlier. To digress into the history of phenylketonuria, Folling's original cases had previously been classed as schizophrenic. The Children's Bureau (1964) reports that because of their behavior, phenylketonurics had often been mis-diagnosed as having early childhood schizophrenia, and lists five cases which were reported as autistic. Woolley has evidence to show that serotonin deficiency may cause schizophrenia, and suggests that after the person has passed the period where excess phenylalanine will cause the mental retardation, and the normal diet is resumed, the continued inhibition of serotonin may produce schizophrenia, which may not appear until adulthood. This evidence suggests that a phenylketonuric should remain on a well-controlled diet until sufficient evidence to the contrary is found.

Recent Studies

Much of the research in the literature at present has been focused on reporting the symptoms of phenylketonuria, and to record the response of people placed on the diet. However, most of the people under dietary control at present have already been damaged by the presence of the excess phenylalanine and by-products in the bloodstream. It has only been four years since the mass screening program went into effect in Oregon, although some patients were screened at birth earlier than this. Probably as a result of this,

there is little information in the literature as to what specific problems are being faced by those children who were screened at birth, and who are at present on the low-phenylalanine diet.

In a study done by Berman, Waisman, and Graham (1966) at the University of Wisconsin Medical Center, the I.Q.'s of diet-treated phenylketonuric children were compared with their siblings, and with untreated phenylketonuric children. The tests used were the Stanford-Binet, and the Cattell Infant Intelligence Scale. In the initial test, the scores of the earliest-treated children were 27.5 I.Q. points below the scores of their unaffected siblings. Children who began the diet treatment at later ages had increasingly lower scores. As the children were re-tested at later dates, the diet-treated children showed an increase in I.Q., while their unaffected siblings remained stable. All differences were significant at the .01 level on a t test. The Binet scores were corrected to remove the effects of the age differences, but the investigators were unable to do this with the Cattell scores. However, while the diet-treated phenylketonuric children gained in I.Q., their test scores still remained lower than their siblings', which indicated to the authors that even though these children were on diets, there was still some factor of phenylketonuria operating.

In another test done by Fisch, Gravem, and Feinburg (1966), at the University of Minnesota Hospital, the effect of the phenylalanine

restricted diet upon the growth of the diet-treated children was studied. In this study, they cited several problems facing children on diet therapy. Restriction of the diet "can result in ... cutaneous leisions, hypoglycemia, bone changes, megaloblastic anemia, and death" (p. 3). They also stated that maintenance of correct phenylalanine blood levels was not always possible because of "accidental or intentional deviation from the diet, lack of parental interest, or illness with temperature elevation and resulting protein breakdown" (p. 4). They also stated that as the child grows older, and increases social activities, it is more difficult to maintain the correct phenylalanine level in the blood.

In their testing, they used the Iowa Growth Charts to plot the weight and height gains of the children. They took these measurements each time the child visited the clinic. Once a year, wrist radiographs were taken to determine bone changes. It was found that the mean weight for untreated boys remained near the normal within the limits of $\pm$1 standard deviation of the mean, from two to ten years of age. The mean height was below normal and exceeded 2 times the standard deviation in four instances. Untreated girls between two and eight years of age showed height and weight above the norms for both boys and girls. Untreated children tended to have near hormal height and weight, except that the boys were slightly below normal.

Using a Student's t test, it was found that the group on the diet

had a significantly slower rate of growth.

> In every case observed, the immediate response of the phenylketonuric child to the reduction of phenylalanine in his diet was a sudden loss of weight and a subsequent gradual decrease in his previous rate of growth. After the initial 'adjustment period,' weight and height stabilized and development continued at a significantly lower percentile level.
>
> In cases where the diet was terminated for a variety of reasons, weight increased rapidly and strikingly (p. 8).

In brief, the literature on the phenylketonuric children deals with phenylketonuria on three levels. First, there is the phenylketonuric child who exhibits mental retardation, along with various behavioral symptoms which generally lead to the institutionalization of the child. Then, after the introduction of dietary treatment, the behavioral symptoms disappear, but the mental retardation remains in the child who has already been damaged by the effects of excess phenylalanine and its by-products in the bloodstream. Finally, there are studies on children with phenylketonuria who have been diet-treated from birth, and who show no signs of either the mental retardation, or the behavioral symptoms previously associated with phenylketonuric children.

The major obstacle to investigating problems of families with a phenylketonuric child is the lack of interpretation of possible consequences of the known dietary restrictions and behavioral characteristics of the phenylketonuric child. The major facts concerning the disorder are known, but the problems faced by the families are not known.

Sutherland (1966), gives an insight into some of the problems that might face the phenylketonuric child who is diet-treated from birth, by reporting the child as set apart by:

> ... special food, special blood and urine tests, special visits to the doctor, special watchful and overprotective eye of parents and teacher. The families are ever mindful of their goal, 'his ability to perform.' We therefore create problems in discipline, behavior, language usage, etc. He may well fall short of fulfillment of his intellectual potential because of the problems his regimen creates (p. 524).

In view of these needs, this study was undertaken to provide as much of the existing background as possible, both in terms of phenylketonuric children, and in terms of the information available in areas of so-called "normal" childhood that can be considered "problem" areas, as well as to form a tentative interview schedule.

REVIEW OF LITERATURE

There is information in the literature on the problems faced by children and families where the children are affected by other chronic illnesses, such as diabetes, heart disease, and asthma. Crain, Sussman, and Weil (1966) have done studies on the effects of a diabetic child on marital integration, family interaction, and sibling relationships. Hurlock (1964) cites areas of development that have become problem areas for children and families where the children are affected by chronic illness. However, as yet, there appear to be no studies on problems faced by children who are affected by phenylketonuria, other than those already mentioned, and apparently no studies published on the problems faced by the families of these children.

It seems reasonable to assume that because of the multiple problems faced by phenylketonuric children, and the severity of these problems, that there might also be an increase of problems in areas such as: learning, parent-child interactions, dependency and overprotection, language development, physical development, emotional development, personality development, peer relationships, and family relationships. Each one of these areas will be treated separately, with a following discussion of the relationship of these areas to the phenylketonuric child and his family.

Learning

> Stated most simply, learning is the establishment of a connection or association between a stimulus and a response where, prior to learning, no such association existed (Mussen et al., p. 127).

Stimuli and responses can take many different forms. People tend to learn responses that lead to the reduction of intense stimulation, such as fear, hunger, or physical pain. A reward is an event or action resulting from a response which gratifies a particular need. The reward generally strengthens the response. There may be learned needs, as well as the innate needs mentioned above. The responses which gratify these learned needs lead to learned rewards. There also may be generalization of stimuli which will lead to generalization of responses. This generalization comes with the development of language and increased language ability (Mussen, et al., 1963).

The type of adjustment a child makes to life is dependent on his heredity, coupled with his understanding of, and adjustment to, his environment. If a child does not understand his limitations, whether physical or mental, then he will not understand why people react as they do. Understanding is dependent upon conceptual development. In order for the child to develop new concepts, he must be able to see relationships between new experiences and old ones. He must be able to comprehend underlying meanings, and must be able to reason. Factors that influence conceptual development are the condition of the

sense organs, intelligence, opportunities for learning, types of experiences, sex, and personality. Concepts that are subjective will be more heavily weighted than concepts of an objective nature--such as foods that the child has come in contact with, versus foods that the child has had no experience with. The child develops values in terms of which concepts are valuable to him.

The types of concepts that a child has about himself and others, influence his behavior. Children develop misconceptions when they have received incorrect information, through limited experience, through faulty reasoning, through vivid imaginations, through unrealistic thinking, and misunderstanding of words. Concepts of bodily functions are often erroneous, and depend upon the opportunities he has had to learn about them. Children show little knowledge of the interrelationship of bodily functions.

The infant forms concepts of himself first, and this affects his concept of others. His concepts of self are based upon his perceptions of what others think of him. This perception is often colored by misunderstandings. In order to have a realistic self-concept, the child must be independent of those on whom he has been psychologically dependent. The young child's self-concept is influenced by his relationships with his mother, and with siblings. As the child relates to others outside the home, he gains a secondary self-concept (Hurlock, 1964).

Discussion

It would be interesting to determine what types of stimuli and responses lead to learning in the phenylketonuric child. In the following areas of discussion, certain factors will be mentioned, which could influence the type of learning the child experiences, including his conceptual development concerning his environment, and himself.

Parent-Child Interaction

Maternal Attitudes

Although there are no nerve fibers joining the mother and infant during pregnancy, the mother's emotions may still affect the infant. When the mother is excited, certain physiological reactions occur, and chemicals such as adrenalin are released into the blood stream. Some of these chemicals may pass through the placenta, and affect the unborn child. These may be irritating to the fetus, and create an increase in fetal activity. If the period of emotional stress on the part of the mother is lengthy, this may have an enduring effect on the infant after birth. The infant may exhibit a high activity level (Mussen, et al., 1963).

Maternal attitudes during pregnancy are related to emotional maturity, and past and present experiences. The behavior of the mother may reflect her emotional needs, her feelings about having

this particular child, as well as children in general. Her behavior and attitudes may also be affected by relationships within the family (Breckenridge and Murphy, 1963). Mothers with unfavorable attitudes toward pregnancy tended to show more problems during pregnancy, such as nausea and vomiting. Also, mothers who showed unfavorable attitudes toward pregnancy reported disturbances in infants related to eating, elimination, sleeping and crying (Mussen, et al., 1963).

Parental Concepts

Parents conceptualize the type of child they will have before the child is born. If the real child matches this concept, he will be more easily accepted by the parents, than if he does not. "Many forms of so-called 'problem behavior' in children are merely discrepancies between the behavior of real children and parental concepts of ideal behavior" (Hurlock, 1964, p. 67). Many cases of emotional instability in children result from the fact that the child is not the ideal child that was expected by parents or siblings.

In relation to children such as "blue-babies", parental anxiety carries over from the time of danger to the rest of the child's life and has a "marked influence on the type of social and emotional adjustments the children can make" (Ibid. p. 89). If the mother is unstable and confused, any apparent problem of the child's will only add to the instability and confusion. Parental attitudes surrounding the birth

situation may have greater effects on the development of the child than the physical hazards of birth. Parents are likely to judge a child's progress and behavior in terms of previous children in the family. It is difficult for parents to separate "similarity and normalcy," and "individuality and abnormality" (Ibid. p. 100).

Congenital Differences in Infants

Infants differ at birth in their biologic individuality. This not only affects physical development, and susceptibility to disease, but may affect behavior, also. Some infants, from birth, are responsive to others, while other infants are unresponsive. The effects of the infant's response help determine the kind of relationship which will be formed between the mother and child (Breckenridge and Murphy, 1963). The activity levels of children differ at birth. These activity levels have an effect on the motor development of the child during the preschool years. Sensory thresholds also differ in infants at birth (Mussen, et al., 1963).

Feeding

Among the basic needs of infants is the gratification of the hunger drive. If immediate gratification is not received, tensions mount, and bodily activity is increased greatly. Because of the importance of this drive, it plays a large role in the early learning of the infant.

The feeding situation is important in establishing a relationship between the mother and infant. Nursing automatically establishes bodily contact between the mother and infant, which may be pleasurable to both. However, body contact can be maintained with bottle feeding, also. The type of contact between the mother and infant is important, as the mother's emotional feelings will be communicated to the infant in the body contact (Mussen, et al., 1963). It is possible that the breast-fed infant may have an advantage over bottle-fed infants in the area of physiological immunity that is contributed by the mother's milk (Breckenridge and Murphy, 1963).

The schedule of infant feeding can be strictly regulated by the mother, or regulated to the demands of the infant. It has been shown that infants can adapt to a regularly scheduled feeding, but that variations in this scheduling produce signs of discomfort in the infant (Mussen, et al., 1963).

According to Breckenridge and Murphy (1963), "An individual's attitude toward food is an expression of his total personality..." (p. 58). A person may be either inflexible in his eating behavior, or may readily accept a variety of foods. Parents influence greatly the behavior of their children toward food. There are emotional factors influencing food habits. Unpleasant experiences associated with food, such as periods of emotional stress, poorly prepared food, and unhappy mealtime situations strongly influence the formation of food

habits. A chronic illness such as diabetes may affect the food habits of an entire family, particularly if the meals are planned around the diet of the individual with the illness (Breckenridge and Murphy, 1963).

Discussion

In terms of maternal attitudes, there is the possibility that the mother of a phenylketonuric child would exhibit a negative attitude toward pregnancy. If the pregnancy involving the phenylketonuric child was her first pregnancy, or if other children in the family were not affected by phenylketonuria, then it would be doubtful that phenylketonuria would have any effect on her attitude, prior to the birth of the child. However, if, as in some instances, older children in the family were phenylketonurics, then it might be reasonable to assume that the mother's attitude toward any ensuing pregnancies might be adversely affected. It would then be reasonable to assume that the infant's behavior following birth would be affected in the manner reported by Mussen, et al.

The parental concept of the child before birth most probably would not match the "real" infant with phenylketonuria, except possibly in cases where there were already children in the family with phenylketonuria, and the parents expected the new child to have phenylketonuria also. In this case, depending on the effects of the diet therapy and its onset, with the infant as well as siblings, parental

concepts could be favorably altered.

It would appear reasonable to assume that there would be a certain amount of parental anxiety surrounding the discovery of phenylketonuria and the subsequent development of the child. Individual differences in the child could add to this anxiety. In terms of individual differences, R. J. Williams goes so far as to say that it is "more important to know what sort of patient has a disease, than to know what sort of disease a patient has" (Williams, 1956, p. 2). Williams suggests that each person has a unique metabolic personality, and because of this, no two people could be expected to react in exactly the same way to the same environment. Following this line of thought, two people with the same disease could react in very different ways to the disease and to their environment. Some of these individual differences might not be related to the phenylketonuria, but it would be difficult to separate those that were.

It would be reasonable to assume that the feeding practices involved in the interactions between the phenylketonuric child and his mother would be very important. The mother would not be able to breast-feed the infant because of the protein content of her milk. The feeding schedule of the infant could also create problem areas. The child would be on a strict schedule and there exists the possibility that hunger demands might not always be met. Also, due to the balance of phenylalanine and by-products in the bloodstream, one could

expect changes in the schedule from time to time, which might cause problems in adjustment on the part of the infant.

It would appear reasonable to assume also that the limitation of variety in the diet, and the strictness of enforcement would have an effect on the child's emotional attitude toward food. This might be influenced adversely and to a greater degree if the child observes other members of the family, and other persons outside the family, eating foods that he is not allowed to have. In turn, the diet of the phenylketonuric child could have an effect on the food habits of the rest of the family.

Dependency and Overprotection

According to Hurlock (1964), children who have a chronic illness often exhibit signs of dependency. They tend to seek help when it is not needed, and seek attention and approval from others. They learn this behavior when too much is done for them, and when they have few opportunities to be independent. In two studies cited by Mussen, et al., (1963), two forms of maternal behavior were associated with high degrees of dependent behavior. These were consistent reward of dependency, and inconsistent reward and punishment.

The consistent reward of dependency may be found in maternal overprotection. This results when the parent encourages infantile care past the period of infancy, and prevents the development of

self-reliance. If the child is considered a helpless infant after the period of infancy is passed, this can deprive the child of the opportunity to master the developmental tasks required of him, in areas of personal and social adjustment (Hurlock, 1964). Overprotection may result in feelings of inadequacy in the child, and may prevent the child from learning enough about his environment to develop feelings of self-confidence. In a study cited by Mussen, et al. (1963), children with overprotective parents were described, in part, as submissive, self-conscious, and suffered from feelings of inferiority and insecurity.

According to Landreth (1967), children naturally seek independence after the first year of life. There may be several reasons for the overportection which would inhibit this independence. One reason may be lack of parental understanding of what the child is capable of. Another may be parental insecurity. If parental nurturance is diminished, and parental control is increased, the children become more dependent. Many of the studies in the literature are concerned with dependency and overprotection in terms of parental permissiveness and strictness. Strictness is associated with dependency, and in particular, where the feeding schedule of the child is controlled excessively by the parent, feeding problems result. In terms of learning, the dependent child is a slower learner, with I.Q.'s of dependent children likely to be lower than those of non-dependent children

(Breckenridge and Murphy, 1963).

Discussion

It would be reasonable to assume that since phenylketonuria can be classed as a chronic illness, that phenylketonuric children might exhibit signs of dependency, and that parents of phenylketonuric children might exhibit signs of overprotectiveness. Because of the fact that the diet must be strictly regulated, and the fact that it would be the duty of the parent to maintain this regulation, the above-mentioned problem in feeding might occur. If, as a result of the diet, the child has additional problems such as hypoglycemia and anemia, as mentioned earlier, this could give rise to parental control not only in the area of diet, but in other activities that the child might normally pursue. As mentioned earlier, the diet must also be regulated in terms of the activity of the child, and in cases of illness of the child. These would add reasons for the parent to exhibit control over the activities of the child. Also, if the children with phenylketonuria have a lower I.Q. than that of their siblings, and if their growth rate is markedly slower, these tendencies could give rise to parental concern and anxiety, which could lead to overprotection and resultant dependency.

Language Development

Language development for the young child is dependent on comprehension, which is in turn, dependent upon understanding. According to Hurlock (1964), the child begins his understanding with the emotional responses of the mother, which being as early as the third month of life. The parents are the major source of speech stimulation in the young child. Their speech, and the responses they give to the young child's speech, determine the child's understanding of the meanings of words. Young children use speech in motor activity, as a means of social identification, and as a means of thinking aloud (Landreth, 1967).

In language development, the child's words increase in specificity and in abstract quality. The two developments proceed simultaneously. The child's ability to use a particular word does not always mean that he understands the meaning of that word in the way that an adult would (Mussen, et al., 1963). The content of a child's speech is a measure of his language growth, his interests, and his personality growth (Breckenridge and Murphy, 1963).

One of the things which may influence a child's growth in language is his mental ability. Children who are retarded generally are retarded in language development, also. Children who receive attention without the need to communicate verbally on their part, are slower in their language development. Children who are limited in

their experiences in general, may be slower in their language development. A lack of practice, both in interaction with adults and with other children, may be responsible for slower development (Breckenridge and Murphy, 1963).

According to Hurlock (1964), illness during the first two years of life may delay the beginning of speech, and of the use of sentences by one to two months. If the child is cut off from play contacts, retardation of speech is more serious. Studies of children's speech show, among other things, the child's concept of himself. It is possible to determine how the child feels about himself and his relationships with others, including his peers. The content of speech also shows the child's emotional state.

Discussion

The studies which have been published, dealing with the speech of phenylketonuric children, are studies which deal only with children who are retarded. In these studies, it was noted that after diet therapy had begun, all symptoms with the exception of retardation, disappeared. It would be reasonable to assume, however, that their language development remained retarded, also.

In children who have been on the diet therapy since birth, there might also be the possibility of language retardation. As has been mentioned earlier, the reported I. Q. levels are lower in these

children than their siblings. This might account for some language retardation. Hurlock's discussion of the effect of illness on language would give added reason to suspect some language retardation. The possibility that the parents might meet the needs of the child without active verbal communication on his part, could also contribute to slower language development. It would be interesting to determine the kinds of verbal concepts the child learns, particularly in terms of his understanding of the nature of his illness. It would appear that at a relatively early age, the parents would need to communicate to the child the reason for his highly restrictive diet, and, in the case of other complicating physical factors, the reason for restriction in physical activity. It would appear that there would be the possibility that the phenylketonuric child might have some conceptual understanding that his non-phenylketonuric age-mates would not have.

Physical Development

At birth, the weight of the infant generally falls between $5\frac{1}{2}$ and $9\frac{1}{2}$ pounds, with a weight loss in the first few days of life. The growth rate in the first two years of life is rapid, but decelerating. In the next year, the deceleration rate slows, and in the remainder of the preschool period, the rate accelerates. Factors which contribute to weight differences in children are heredity, and environment, particularly in the area of nutrition. Also, individual children fluctuate

in their weight growth.

Skeletal growth proceeds from cartilaginous tissue formed before birth. Ossification begins around the middle of the sixth week after fertilization, and is not generally completed until the individual is in his twenties. In some instances, where there has been severe illness, or malnutrition, bone scars may appear. Chronic malnutrition may retard skeletal maturation.

The infant is born with all of the muscle fibers he will have, but throughout the developmental period, there is an increase in growth and maturation of these fibers. Children with less dense muscle tissue have been shown to have both muscular and mental fatigue. The condition of the muscles is dependent upon the health and nutrition of the child (Breckenridge and Murphy, 1963).

"The child's physical development has a marked influence on the quality and quantity of his behavior" (Hurlock, 1964, p. 111). This may affect directly what he is able to do, as well as indirectly, in what he perceives himself as able to do, and how he perceives other's attitudes toward him. Any disturbance in the normal growth or functioning of the body upsets the homeostasis of the body. The longer the duration of this disturbance, the greater the effect on the child's behavior.

The importance of body size in relation to the psychology of the child is dependent upon how others, peers in particular, react to his

size. Illness of any kind affects the child in the areas of development, behavior, attitudes and personality. The child's attitude toward the illness is determined by the attitudes of significant people around him. Prolonged illness affects the child's status in his social group, particularly if he has to be protected. If he is removed from this group, he fails to learn patterns of social behavior. He may develop unhealthy attitudes about his own importance. "Illness of any kind limits the child's activities" (Hurlock, 1964, p. 146).

Discussion

In the area of physical development, it would appear that the phenylketonuric child would encounter several difficulties. In the study by Fisch, et al. (1966), the children with phenylketonuria who were on diet therapy showed slower rates of growth than normal, as judged by the Iowa Growth Chart. It would appear that the phenylketonuric child would be smaller than his "normal" age-mates. From the discussion by Hurlock, this could affect the way in which he is accepted by his peers. It could also affect parental attitudes, as brought out earlier.

Since the phenylketonuric child is on a protein-restricted diet, it would be reasonable to assume that skeletal and muscle growth would be affected, since protein intake is largely responsible for growth in these areas. One could expect slow muscle and skeletal

maturation, as well as accompanying symptoms such as fatigue. This in turn, would affect the child's physical activities, which would affect his social activities. Difficulty in these areas could be expected to affect the emotional development of the child.

Emotional Development--Fear, Anxiety, and Anger

According to Breckenridge and Murphy (1963), the earliest emotion in infancy is described as non-specific. As the child grows, his emotional behavior changes in accordance with his constitution and environment. His emotional responses differentiate into responses of fear, anger, and affection. More complex emotions such as guilt, sadness, joy, anxiety, and disgust, develop later, as the child increases his comprehension of his environment (Mussen, et al., 1963).

The fears of a young child are generally in response to specific things such as noise and strangeness. As the child grows older, his fears take on a more imaginary aspect (Breckenridge and Murphy, 1963). Specific fears depend upon the child's intellectual maturity. An association of a present experience with a past experience which was unpleasant can produce fear. Association and identification with a fearful person can stimulate the child's learning of fear (Mussen, et al., 1963). The child's health is a predisposing factor in the onset of fear (Landreth, 1967).

There will be variations in what children will fear, depending

upon their physical and mental development. Retarded children will have fears characteristic of younger children. As children grow older, they inhibit the impulse to show fear. They may withdraw from a situation that is fear producing, or develop imaginary ills, or show overt signs of fear. Worry is considered the fear of an imaginary situation. Worry often leads to anxiety, which is the fear of an anticipated situation of which the child is not consciously aware. Although anxiety often develops in later childhood, its beginning is in the fear of early childhood (Hurlock, 1964).

Anxiety may result from fear of potential physical harm, and from fear of loss of love of either parent or peer. It may result from the child's insecurity in his ability to master his environment. It may result from a false self-concept (Mussen, et al., 1963). Anxiety may result from basic needs not being met (Breckenridge and Murphy, 1963). Common worry leading to anxiety often centers around the child's health (Hurlock, 1964).

The child's ability to withstand anger-producing stimuli depends upon his physical and emotional condition. In a study reported by Landreth (1967), children who were ill or tired showed more outbursts of anger than when in better physiological condition. Situations that give rise to anger include blocking of activities, thwarting of wishes, and cumulative irritations. Preschool children often object to having to do what they are told to do. Anger responses are

often violent outbursts of temper. Impulsive expressions of anger are more common in younger children, and appear before tne inhibited expressions such as withdrawl.

The "status of the child in the family is an important factor in determining how the child will express his anger" (Hurlock, 1964, p. 287). Children who show the greatest need for affection and attention often express their anger in the inhibited form. The child who is subjected to authoritarian child training will often develop a hostile attitude toward all people in authority (Hurlock, 1964).

> Whenever homeostasis is upset, owing to fatigue, poor health, or developmental changes in the body, the child experiences emotionality.
>
> A low energy level resulting from malnutrition, for example, makes a child irritable, depressed, unsocial and reserved. Children who have a history of illness are more emotionally unstable than those whose health is better (Hurlock, 1964, p. 302).
>
> Any chronic disturbance, such as asthma or diabetes, produces more or less constant emotional tension (Ibid., p. 303).

There are indications that children with lower intellectual levels have less emotional control. Parental attitudes may affect the child's emotional state. If the parent is overanxious, this is communicated to the child. Also, if the parent constantly talks about the child's problems, or makes the child the center of life at home, the child's emotional problems will increase.

If the child experiences frequent periods of heightened emotionality, these can upset the homeostatic balance of the body, and disturb

its normal functioning. This can also result in decreased mental efficiency (Hurlock, 1964).

Discussion

It would be reasonable to assume that in the life of the phenylketonuric child, there would be several situations which could be fear-producing, and lead to the development of anxiety. According to the above discussion, the child's illness could cause him to be physiologically more susceptible to fear producing stimuli. Frequent visits to the doctor, and continuous physiological and psychological testing situations could be fear-producing stimuli. The possible anxiety over the illness of the child on the part of the parents, could be communicated to the child, and thus create feelings of anxiety in the child. The child's illness could affect his interaction with his environment, and his self-concept, in a negative manner, and create anxiety about his ability to cope with the environment. Because of illness, the child would possibly be more prone to show anger, due to restraint of activities, and thwarting of wishes.

It would appear reasonable to assume that the phenylketonuric child would exhibit the same kinds of emotional instability shown by children with chronic illnesses, as described by Hurlock. Also, if the diet-treated phenylketonuric child maintains a lower intellectual level, less emotional control could be expected. The way in which

the child is treated, as a member of the family, could also affect his emotionality. If parental concern is over the phenylketonuric child, to the exclusion of other members of the family, his emotional stability would be affected.

Personality Development

> Personality development may be viewed as a continuous, highly complex process, involving the interaction of a biological organism with its physical, psychological, and social environment (Mussen, et al., 1963, p. 5).

All of the aspects of personality development are interrelated, with the emotional health of the child having the ability to affect the physical health of the child, and vice versa (Mussen, et al., 1963). There are basic, inherited individual differences in the personalities of children, with some tending to be more stable, physiologically, psychologically, and socially, while others tend to be unstable in these areas. Neither of these qualities is more acceptable than the other (Breckenridge and Murphy, 1963).

Given the basic, inherited personality, the interraction of this personality with the environment determines the direction personality development will take. Chronic illness would be a part of the child's inherited personality that would affect his total environment. Hurlock (1964) discusses some of the problem areas that affect the personality development of the child with a chronic illness. This child will tend to expect the same treatment from people outside the home as that he

receives at home. Children who are ill may have low energy levels, and as a result, become shy, reserved, irritable, depressed, and unsocial. Intelligence levels affect the personality, with the lower levels being adversely affected. Children with too many worries often lack self-confidence, and develop a lower frustration threshold.

In studies with children who suffer from asthma and those suffering from cardiac conditions, the same personality syndrome was found.

> This led to the conclusion that children suffering from any chronic illness, whether physical or psychological in origin, 'display an emotional pattern that deviates from the normal' (Hurlock, 1964, p. 147).

Children suffering from diabetes experience tension and anxiety and a resulting frustration which often leads to aggression.

If there is a problem with diet, and the mother's attitudes cause guilt in relation to eating, there will be an effect on the personality. Self-acceptance is aided by the child's demands for himself being kept within his level of achievement. A child who is handicapped must be able to realize that while there are things he cannot do, there are also things that he can do. The child must also be able to adjust to how others see him in relation to how he sees himself. His self-acceptance is influenced by the stability of his self-concept (Hurlock, 1964).

Discussion

In terms of the child's expectations of treatment outside the home in relation to the treatment within the home, the view of Sutherland, et al. (1966), of the phenylketonuric child as "special," gives some indication of what the child's expectations might be. Because of the restrictive diet, and complications brought about by the diet, it would be reasonable to assume that the child would have a lowered energy level, leading to the personality problems suggested by Hurlock. In view of the reported problems of the phenylketonuric child, such as slower growth rate, and lower I.Q., it would also be reasonable to assume that the child's self-concept and self-acceptance would be adversely affected.

Peer Relationships

Although peers do not play an important role during the first three years of life, they become increasingly important from that time. As brought out earlier, the expectations of the preschool child in social areas outside the home are determined by the experiences within the home. The behavior learned in the home is not fixed behavior, and may be modified by the peer group. In social participation, the child's behavior will be modified by experience with rewards and punishment for particular social responses.

Children who are dependent on adults for support will experience difficulty in establishing relationships with peers. Both Mussen, et al. (1963), and Swift (1964) report that the dependence on adults is negatively correlated with social acceptance by peers.

Although preschool children may tend to form friendships with children of the same sex, a criterion for friendship listed for both sexes was similarity in physical ability. Children of this age tend to find social acceptance in copying the behavior of other children of like sex (Mussen, et al., 1963). Hurlock (1964) reports that children with physical handicaps often are able to participate only in activities with low prestige value. Also, children who have lower mental ability have poor social acceptance.

Discussion

If, in fact, the phenylketonuric child does exhibit any or all of the problems mentioned previously in this paper, such as dependency, emotional problems, retardation in areas of physical development, mental ability and language, etc., it would be reasonable to assume that he would also experience difficulty in forming peer relationships.

Family Relationships

Whenever the accustomed pattern of life in the home is changed, adjustments must be made by all members of the family. If not,

trouble may result. Once poor relationships are established, they often persist, and grow worse. Behavior and attitudes of the parents may create problems within the family. One such behavior already discussed is overprotectiveness. Another behavior is permissiveness, which may result in indulgence. Covert rejection of one or more children on the part of the parents can create problems. Other problem areas are submission to the child on the part of the parents, which leads to the child's domination of the home, and favoritism shown by the parent toward one child. Favoritism on the part of the parent can lead to jealousy and resentment of the favored child by the siblings (Hurlock, 1964).

Although, according to Hurlock (1964), sibling relationships are influenced by the ordinal position of the child in the family, by age differences, and sex of siblings, Farber (1959) reports that a retarded child is regarded as youngest in the family, regardless of birth order. In studies with diabetic children, Crain, et al. (1966b), found that the illness affected the other children in the family in that the mother formed closer ties with the diabetic sibling.

In a study reported by Farber (1959), parents who see the child's illness as coming from a source other than themselves are less likely to be adversely affected than those who see a relationship between themselves and the illness. In observations of families with retarded children, Farber (1964) noted that these families tend to

participate less in community activities. Care of the child is involved in this, as well as the fact that the demands on the family because of the illness may prevent "normal" participation. He defines a crisis as "any decisive change which creates a situation for which the habitual behavior patterns of a person or group are inadequate" (1964, p. 390). He goes on to say that in a family with a handicapped child, the existence of a crisis depends on the extent to which this changes the family situation. The handicapped child presents the potentiality of a crisis situation. Again, in studies with families of diabetic children, Crain, et al. (1966a), found an association between the presence of a diabetic child in the family, and lower marital integration and greater marital conflict among parents.

Discussion

It would appear reasonable to assume that the presence of a child with phenylketonuria would change the accustomed pattern of the home situation. Whether or not this would result in a crisis situation, as described by Farber, would depend on the quality and quantity of the problems presented by the child. Although the diet-treated child is reported as being lower in I.Q. than his non-phenylketonuric siblings, it is questionable whether he could be considered "retarded" in the extreme sense.

The relationships of the phenylketonuric child with his siblings

could possibly be affected in terms of the discussion by Hurlock, and the findings of Crain, et al. There exists the possibility that the phenylketonuric child would have closer ties with the mother, as well as being the object of favoritism on the part of the parents.

Since phenylketonuria is genetically inherited, it would appear reasonable to assume that the parents might feel responsible for the child's illness. Whether or not the illness of the child would affect the family participation outside the home would be dependent on the amount of care required by the child, and the family adjustment to the illness.

Family Developmental Tasks

The need for a conceptual framework for the study of the family has been felt for some time. One of the recent attempts to provide this conceptual framework has been the development of the concept of family developmental tasks. As yet, this concept is too general to be studied in depth. However, it is interesting from the standpoint that it has provoked a great deal of thought and controversy in recent years. It is possible that in time, the concept of family developmental tasks will reach the stage where it can be empirically verified, and therefore, the effect of the phenylketonuric child on the developmental tasks of the family, and of the child, himself, is well worth considering.

The strongest support for the concept of developmental tasks is available in the literature describing the prenatal phases of development. During this period, the development of each organ proceeds in an orderly sequence. If interruption occurs at any particular point of the sequence, for example, during the stage which is devoted to the development of the eye, the development of the eye will be arrested. Complete development during the prenatal period depends not on the nature of the interruption, but upon the timing of the interruption. If any stage does not develop at the appropriate time, the next stage will begin, and the priority will go to that stage, with the previous one never being able to develop fully (Erikson, 1963).

The concept of prenatal development occurring in an exact, orderly sequence was then carried over to the development of the individual after birth, and throughout the life cycle of the individual. This was probably first defined by Havighurst (1965), as follows:

> A developmental task is a task which arises at or about a certain period in the life of an individual, successful completion of which leads to his happiness and to success in later tasks, while failure leads to unhappiness in the individual, disapproval by society, and difficulty with later tasks (p. 2).

Hill and Rodgers (1964) describe, in detail, the efforts made since the 1930's to carry this concept into the family setting. The concept of family developmental tasks has formed the basis of texts by Duvall (1967), and Stroup (1966). Duvall sees the family as being composed of individuals, each with their separate developmental

tasks, with the addition of the developmental tasks that arise because of the nature of the family. She sees the interdependence of family members as being important, as the role and tasks of one member may depend on the role and tasks of another.

Duvall paraphrases Havighurst by stating:

> A family developmental task is a growth responsibility that arises at a certain stage in the life of a family, successful achievement of which leads to satisfaction, and success with later tasks, while failure leads to unhappiness in the family, disapproval by society, and difficulty with later developmental tasks (p. 45).

One of the settings in which the developmental task concept is seen is that of the family life cycle. The stages of the cycle, depending on the author, can number from four to seven. Stroup (1966) lists the following:

1. The Childless Couple
2. The Expectant Pair
3. The Pre-school Family
4. The School-age Family
5. The Teen-age Family
6. The Family in the Middle Years
7. The Family in Later Years.

The stages may vary in time of occurrence from family to family. One family may remain in the first stage for several years, or even a lifetime, while another may find themselves in the second stage immediately after marriage. A family which has reached the fifth

stage may find themselves again in the second stage, and their reactions to being in the second stage would be expected to differ from the first time they were an expectant pair, and from other couples who are going through this stage for the first time.

The developmental tasks of the family which occur throughout these stages also differ according to author, slightly, and in order of importance, but in general, remain basically the same. They are not necessarily expected to occur at any given stage in the family life cycle, nor is their occurrence expected to be the same in any two families. The developmental tasks which are basic, and which occur throughout the life cycle are listed by Stroup (1966) as follows:

1. Maintenance of order and intra-family stability.
    a. Providing mechanisms to facilitate channeling of emotional ties among family members.
    b. Controlling sex expression.
    c. Establishing channels of communication in the family.
2. Establishing a working relationship with the larger family network and the community.
    a. Developing a balanced relationship between intra-family living and relations with the outside world.
3. Providing a physical base for family members.
    a. Providing clothing, food, and housing.
    b. Providing an economic undergirding for the family.

4. Dividing and allotting responsibility and authority.
    a. Providing a means of determining who does what, when, and why.
    b. Organizing and dividing the work of the home.
    c. Developing habitual roles (reciprocal) so that tasks are performed on a reasonable schedule.
5. Developing a family value system and pattern of socialization
    a. Developing a system of values and goals on which there is a basic agreement among adult members.
    b. Developing a philosophy and method of child-rearing.
    c. Socialization of children into increasingly more mature roles.
6. Maintenance of morale and incentive.
    a. Developing patterns to give recognition and reward to family members achieving family goals.
    b. Developing emotional release mechanisms that are reintegrative for family structure.
    c. Providing a systematic method of assuring support to family members needing aid and encouragement.
7. Regulating reproductive processes.
    a. Mastering negative and positive control of conception.
8. Development of techniques of incorporating new members into the family, and releasing the young at the appropriate

time.

a. Assimilating new members (by adoption or from the wider family circle) into the family group and home.

b. Releasing members at young adulthood to marriage and the larger social and economic world (p. 66-67).

Some of the developmental tasks of the adults in the family center around providing for the developmental tasks of the children. Others center on the adults themselves, and others center on the family as a whole. The manner with which the family meets these needs will vary according to the need and the stage of the family in the life cycle. Breckenridge and Murphy (1963) indicate that the successful completion of individual tasks for each family member helps promote the successful completion of the tasks of the family as a whole. In accordance with the definition of family developmental tasks, as given by Duvall (1967), they elaborate on the idea that the successful completion of these family tasks at any particular stage in the life cycle helps to promote the successful completion of tasks at later stages in the life cycle.

According to authors such as Stroup (1966), and Duvall (1967), the tasks of the young couple center first on their adjustments to each other, and to the marriage situation. Then the tasks move to the adjustments of pregnancy and to the birth and acceptance of a child. It is the tasks in these latter categories which are of most concern.

They need to develop emotional schemes which will take care of the growing family. They need to establish patterns of emotional attachments, with the three important factors being love, demonstration of affection, and emotional interdependence. Adjustments to these factors will, of course, continue throughout the marriage.

In the midst of the resolution of the tasks of early marriage often comes the task of facing and accepting pregnancy. The problems of marriage are usually complicated by the arrival of a child. Some of the tasks developed, such as division of roles and responsibilities may change, and other tasks, such as the development of an adequate philosophy of life, take on new importance. There may be problems such as changing the living conditions to make a place for the baby. Also to be considered are the new strains on the family income.

After the birth of the child, the parents now encounter the task of being responsible for the life of another human being. Since the suggestion has been made that parents do need to learn to love their children, one of the important tasks facing the couple is that of learning to love the infant. Common routines are reorganized around the child, and the parents then become involved in helping the child with his individual developmental tasks, such as: adapting to his environment, giving and receiving affection, developing a sex role, learning to communicate, learning the family value system, eating, toilet habits, developing motor skills, learning how to cope with hazardous

situations, developing autonomy and independence, learning to handle emotions and impulses, learning how to react to frustration, and many others.

Other tasks in the life of the couple often need re-evaluation and readjustment at this point, and as the family expands, new tasks take on importance as a result of the addition of children. Some of these tasks are: providing for expansion of monetary needs, providing adequate space and time for each individual, providing for the socialization of the children through exposure to the outside world, development of new patterns of responsibility which include the children, providing for the changes in the emotional constellation of the family, and maintenance of morale and motivation to carry out family tasks (Stroup, 1966; and Duvall, 1967).

The Study of Family Developmental Tasks

Hill and Rodgers (1964) express the viewpoint that longitudinal studies would be best for studying the developmental task concept, and suggest that "deviant" families might be a good direction for study. A "deviant" family would be one in which a member, such as the father, was absent, or one in which there was a problem such as a chronic illness.

The previously discussed developmental tasks are generally discussed in terms of families in which both parents are present, and all

members in relatively good health. It might therefore be fruitful to investigate the effects of the birth of a child with phenylketonuria upon the developmental tasks of the family. It would be reasonable to assume that any of the problems concerning the phenylketonuric child and his family, which are previously discussed in this paper, would have an impact on the family developmental tasks because of the fact that developmental tasks are involved in the relationships between children and parents; between siblings; and between the entire family and the wider circle of associations outside the home. The strict diet of the phenylketonuric child, alone, could be expected to affect not only the developmental tasks of the child, but also those of the entire family.

Some of the tasks which could be expected to be affected are: maintenance of order and intra-family stability, establishing a working relationship with the larger family network and the community, dividing and allotting responsibility and authority, developing a family value system and pattern of socialization, maintenance of morale and incentive, and regulation of reproductive processes. Just as the list of developmental tasks itself is not exhaustive, either for the family or for the individual, the list of developmental tasks which could be affected by the presence of a phenylketonuric child in the family is not exhaustive, nor will any attempt be made at this point to determine the direction and intensity of the effect upon the developmental tasks.

Down's Syndrome or Mongolism

One area of the literature which was considered as a source of information on children with mental retardation or chronic illness was that of children with Down's syndrome or mongolism. After a review of studies such as those by Silverstein (1964), Appleton and Pritham (1963), Shotwell and Shipe (1964), McIntire, et al. (1965), Ray, et al. (1963), and others, it was determined that the studies on these children were comparable to those done on phenylketonuric children prior to the introduction of the diet. The studies related to the family were those concerned with the difference between institutional rearing of the child and home rearing. It therefore appears that this area of the literature is not applicable to the present concerns of this study.

METHOD

In an exploratory study of this nature, one of the more useful methods of gaining insights into the possible existence of problem areas within the families of phenylketonuric children would be the "focused interview" as described by Selltiz, et al. (1966). After careful study of the problem area, the interviewer would know in advance the topics and aspects he would wish to cover, and could focus attention on these areas in the interview. It would appear that open-ended questions in the initial interviews would present the possibility of the interviewer gaining additional insights other than the possibilities already considered.

One of the approaches to the study could be by using an initial interview session for the purpose of determining the more fruitful areas of study, to be followed by additional studies using control groups. Another possibility would be to use three groups, for purposes of control, in the initial interview session, with one group being the families of phenylketonuric children, and another group of families with children who have a chronic illness other than phenylketonuria, and a third group of families with children who do not have a chronic illness. The study would focus on families with the children in question being five years of age and younger. Also, the use of a tape-recorder during the interview sessions would provide for objective recording of the responses given.

Interview Schedule

The sources of information leading to the tentative formulation of the questions making up the interview structure fall into four categories. The first set of questions is for the purpose of obtaining general information about the family. The second set of questions stems directly from the literature dealing with the restrictive diet of the phenylketonuric child. The third set of questions is derived from the normative-descriptive data on childhood problem areas, as covered in the review of literature, that could be heightened by the child having a chronic illness such as phenylketonuria. The fourth set of questions is inferred from the normative-descriptive literature, and is an attempt to determine the effects of the phenylketonuric child on the family.

I. General Information.

A. Age of parents.

B. Length of time married.

C. Family Size.

D. Ages of children. } Ordinal position in family.

E. Sex of children. }

F. Age and sex of phenylketonuric child.

G. Occupation of parents and source of income.

H. Educational background of parents.

I. Other persons living in the home.

J. Sources of help in dealing with the child.

1. Those sources known to the family.

2. Those sources used by the family.

II. Questions Relating to the Diet.

A. What form did the diet take in infancy?

Were there any problems associated with the feeding schedule in infancy?

When did the child first begin to eat solid foods?

What kind of solid food was first introduced to the infant?

How did the infant accept this food?

B. Since the child is on a special diet, does this create any particular problems?

Could you describe these problems?

How strict is the diet?

Does the child attempt to deviate from the diet?

Does the diet of the child have any effect on the meal preparations for the rest of the family?

How does the diet of the child affect participation in eating situations outside the home?

C. Do common childhood illnesses create any special problems?

If so, what are these problems?

D. What are the activities that the child normally engages in?

Are there any activities that the child cannot engage in?

If so, what are the reasons for his restrictions?

Does the child participate in play activities outside the home, or primarily within the home.

Is the child physically able to compete with his playmates and siblings?

III. Questions Derived from Normative-Descriptive Data.

A. Would you describe the child as shy and reserved, or outgoing?

If the child is reserved, are there particular situations in which the child exhibits this behavior?

B. How would you describe the child as an infant?

Was the infant generally active, or quiet?

Some infants are more responsive to attention than others, in that some like to be cuddled, while others do not. How would you describe your child, as an infant, in these terms?

In areas other than the diet, do you see any of your child's behavior as an infant, as being related directly to phenylketonuria?

C. Do you notice any problems in the area of language development?

How does the child compare with his siblings in language development?

Does the child understand the nature of his illness?

How has this been explained to him?

D. What are some of the fears that the child expresses?

How does the child express anger?

What types of situations cause the child to express anger?

IV. Questions Relating to the Effect of the Child on the Family.

A. Would you describe the pregnancy prior to the birth of this child as being the same as previous pregnancies?

If not, how did it differ?

(If there has been a previous occurrence of phenylketonuria in the family, or if there are children in the family younger than the child with phenylketonuria) Did you experience feelings of fear during the following pregnancy that the child might also have phenylketonuria?

B. What effects do you see within the family as a result of having a child with phenylketonuria, other than those related to the diet?

What activities outside the family does your family normally participate in?

Do you see any effects on social relations outside the family as a result of having a child with phenylketonuria?

If so, how would you describe these effects?

C. Judging from your experiences with a child who has

phenylketonuria, and so that others who give birth to children with phenylketonuria might benefit from your experience, is there any information that you feel would have been helpful to know in the beginning?

DISCUSSION OF POSSIBLE RESPONSES TO INTERVIEW SCHEDULE

If any of the suggested problem areas in this study do, in fact, turn out to be problem areas in families with phenylketonuric children, there are some suggestions on the basis of existing literature, which might be of help to the families in their adjustment to these problems.

To begin with, it might be helpful for the parents to know that some of the problems they face could also be problems in "normal" families. Children who do not have phenylketonuria could have problems resulting from overdependence on the parents. Parents of children who do not have phenylketonuria could feel insecure in their parental roles. Children who do not have phenylketonuria could be slower than other children in their language development, and in physical development. Children who do not have phenylketonuria could have emotional problems. Children who do not have phenylketonuria could have problems in social adjustment both within the family and with others outside the family.

It might be helpful for the parents to be aware of some of the factors which lead to problems in these areas, particularly in terms of children who do have a chronic illness. An understanding of some of the factors which predispose dependency in the child might aid the parents in helping the child become more independent. An understanding of the effects of illness on language development as well as

an understanding of how language development proceeds in general, might aid the parents in helping the phenylketonuric child increase his language ability.

In terms of the physical development of the child, and his adjustments to any limitations placed on him because of this, it might be helpful for the parent to understand how development normally proceeds, and what the dietary effects on this development are. This might then help them in understanding the growth of their child, and hopefully, they could then help the child, as suggested by Hurlock, in understanding that while there are things that he cannot do, there are things that he can do.

It might be helpful for the parents to understand the factors that cause fear and anxiety in childhood. They might then be alert to things in their child's environment which could be fear-producing, and thus help to reduce these fears.

An awareness of the factors which lead to emotional problems, particularly in terms of the phenylketonuric child, might aid the parents in coping with these problems. An understanding that the effects of the diet and the disease on the physical constitution of the child affect areas such as his frustration threshold, and his ability to control his emotions, could be helpful in these areas.

Above all, an understanding of the individual differences in people might help the parents in relating to their child as a child who

is an individual, rather than a child with a special problem. The parents may see a difference between the child with phenylketonuria and other children, but if they were to compare children without phenylketonuria with each other, they would find differences in these children, also. The child with phenylketonuria does have special problems that other children do not have, but it would appear beneficial to both the child and the family, if the special problems could be treated as individual differences, rather than problems.

Maple-Syrup Urine Disease and Galactosemia

Two other relatively prominent inborn errors of metabolism causing mental retardation, which are screened by the Guthrie test, are maple-syrup urine disease and galactosemia. Like phenylketonuria, both involve the lack of an enzyme in the body which will convert a particular compound, in the food that we normally eat, into the required product for use in the body. As with phenylalanine and its by-products, these also build up in the bloodstream, causing damage to the brain. The present method of averting mental retardation, in all three cases, is through strict control of the diet.

Because of the low incidence of maple-syrup urine disease and galactosemia in Oregon, it appeared more profitable to consider phenylketonuria, alone. It would be reasonable to assume that the problems which might face the phenylketonuric child and his family,

could also face families with children who have maple-syrup urine disease and galactosemia. Therefore, the concerns of this paper could apply to these other inborn errors of metabolism, as well as to phenylketonuria.

SUMMARY

This study was intended to be an exploratory study. As described by Selltiz, et al. (1966), the broad purposes of an exploratory study are,

> ...to gain familiarity with a phenomenon or to achieve new insights into it, often in order to formulate a more precise research problem or to develop hypotheses (p. 50).

The purpose of this study originally, was to gain familiarity with, and insights into, family reactions and adjustments to the birth and subsequent rearing of a child with the inborn error of metabolism, phenylketonuria. In general, the study was to focus on families with children five years of age and younger.

It was hoped that on the basis of this work, a more precise research problem could be formulated. It was also hoped that eventually as a result of efforts in this area, information could be provided for parents of phenylketonuric children which would help them adjust to the problems they might face.

According to Selltiz, et al. (1966), one of the methods of generating useful hypotheses in an exploratory study is the survey of people who have had practical experience in the field of interest related to the study. In the area of phenylketonuria, persons with practical experience would include the medical personnel who have worked with the children. It would be reasonable to assume that through the medical personnel, information on phenylketonuric children would be

available in areas such as intellectual and physical development, previous history of phenylketonuria in the family, the dietary regimen of the children, and problems encountered in the area of the diet. Information from medical personnel could also give possible insights into problem areas not already considered. On the basis of information obtained in these areas, more specific and useful questions for interviewing the families involved could be formulated.

The original method for carrying out this study was to obtain existing medical information on the phenylketonuric children in Oregon, who have been diet-treated from birth, and then to interview the parents of these children in hopes of gaining insights into the above mentioned areas. However, due to complications, this was not possible.

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APPENDIX

The following diagram has been prepared to help clarify the discussion of the pathways involved in the metabolism of phenylalanine.

When the protein is taken into the body, it is broken down into its primary components, amino acids. Three of these are significant in phenylketonuria. They are: phenylalanine, tyrosine, and tryptophan. Part of the tyrosine is converted, with the help of an enzyme, into melanin. Part of the phenylalanine is converted with the help of an enzyme, into tyrosine. Part of the tryptophan is converted, with the help of an enzyme, into serotonin.

In phenylketonuria, the enzyme system for the conversion of phenylalanine into tyrosine is missing. Therefore, phenylalanine is converted into phenyllactic acid, phenylacetic acid, and phenylpyruvic acid. These build up in the bloodstream and inhibit the enzyme which converts tryptophan into serotonin. At the same time, because of the lack of the enzyme between phenylalanine and tyrosine, there is less tyrosine to be converted into melanin.