

NEUROPHYCHIATRIC DISORDERS OF
BLOOD DISEASES IN CHILDREN

Essay

SUBMITTED FOR PARTIAL FULFILMENT FOR
MASTER DEGREE IN NEUROPSYCHIATRY

BY

MOATAZ MOHAMED MANSOUR ELSAFTY

M.B.B.Ch.

618.928
M.M



SUPERVISORS

* PROF.DR.SAMEHA ABDEL MONEIM
Prof. of Neurology.
Faculty of Medicine
Ain Shams University.

* PROF. DR. ZEINAB BISHRY
Prof. of Psychiatry
Faculty of Medicine
Ain Shams University.

FACULTY OF MEDICINE
AIN SHAMS UNIVERSITY

(1985)



C O N T E N T S

	<u>PAGE</u>
- INTRODUCTION	i-ii
- PSYCHOLOGICAL DEVELOPMENT OF HEALTHY CHILDREN...	1
- PSYCHOLOGICAL SEQUELAE OF CHRONIC CONDITIONS....	5
- DEVELOPMENT OF THE CHILDREN WITH CHRONIC ILLNESS	8
- PSYCHOLOGICAL EFFECTS OF HOSPITALIZATION.....	15
- CLASSIFICATION OF BLOOD DISEASES.....	19
- ANEMIA	
. Neurological Manifestations of Anemia in General	31
. Psychological Manifestations of Anemia in General	32
. Pernicious Anemia	34
. Folate Deficiency Anemia	38
. Iron Deficiency Anemia	42
- HEMOLYTIC DISORDERS	
. Sick cell Diseases	49
. Thalassemias	59
. Hemolytic Anemia	65
. Acanthocytosis	67
- POLYCYTHEMIA	70
- PORPHYRIA	77
- COAGULATIVE DISORDERS	79
. Hemophilia	79
. Thrombotic Thrombocytopenic purpura	89
- DISORDERS CHARACTERIZED BY ADENOPATHY, SPLENOMEG- ALY AND/OR ABNORMALITIES OF LEUCOCYTES OR IMMUN- OGLOBULINS	96
. Infectious Mononucleosis.....	96
- NEOPLASTIC DISORDERS	
. Neurological Manifestations	100
. Psychological Manifestations	120
- DISCUSSION	133
- S U M M A R Y	151
- REFERENCES	153



ACKNOWLEDGEMENT

Firstly, ultimated thanks to ALLAH ..

I would like to express my deepest gratitudes to PROF. DR. MAHMOUD MOSTAFA, Professor and Head of Neuropsychiatry Department, Faculty of Medicine, Ain Shams University, for having given me the chance to undertake the subject of this essay.

I would like to thank my eminent PROF. DR.SAMEHA ABDEL MENEIM, Professor of Neurology, Faculty of Medicine, Ain Shams University for her continous guidance, help, advices, and meticulous revision of every possible details of this essay, and also for her efficient supervision,patience and cooperation.

I am also greatly indebted to PROF. DR. ZEINAB BISHRY, Professor of Psychiatry, Faculty of Medicine, Ain Shams University, for her encouragment, profuse sympathy, constant support, constructive criticism and everblasting patient. She ungrudgingly devoted a lot of valuable time in helping me with so many references and without her help, my work could not have been satisfactory accomplished.

I would like to take the opportunity to thank all my Professors, and colleagues especially DR. MOHAMED REFAAT EL FIKY, Assistant lecturer in Neuropsychiatric Department, Ain Shams University, among whom I felt sincerely and kindness and from whom I have received great help, cooperation, and encouragment.

* * *

MOATAZ EL SAFTY
1985

INTRODUCTION

Blood diseases in children are mostly considered to be chronic conditions. The effects of chronic condition on normal growth and development of the children and the family life pattern are pervasive.

Blood diseases of children are present with various neuropsychiatric disorders depending on the nature and severity of the diseases. The central nervous system, spinal cord and cranial and peripheral nerves, are the main parts in the body which may be affected resulting in various signs and symptoms.

Anemias cause irritability, lack of interest in surrounding narrow attention spans, perceptual restriction, pagophagia, trichophagia and trichotillo mania. (Weitzman 1984) Neurologically, it may cause neurologic pain, numbness, tingling and rarely may cause clinical picture of pseudo-tumour cerebri (Merin, et al., 1968).

Hemorrhagic blood diseases may cause intra-cranial, intraspinal hemorrhages and cranial and peripheral nerve lesions (SlutschHg, et al., 1981). Psychologically, the behaviour of the child is referred to as risk-taking. The child may recognize himself as a disabled child leading to weak, undeveloped ego and sense of inferiority resulting in social isolation (Agle, 1974).

Diffuse infiltration of the leptomeninges by neoplasms of hematopoietic system, producing what is called "meningeal syndrome", Neuropsychiatric disorders of malignant blood diseases may occur due to the disease itself or due to therapeutic modalities used. (Altman, et al., 1978).

So, the aim of this study is to write a review on neurological and psychiatric disorders complicating blood diseases in children to ensure proper diagnosis and good management of these cases as early as possible to prevent any disability in the future.

❖ ❖ ❖

" TO MY PARENTS "

" REVIEW OF LITERATURE "

PSYCHOLOGICAL DEVELOPMENT OF
HEALTHY CHILDREN

To understand the effect of blood diseases on psychological functions, we have to know the psychological development of healthy children.

The child at all ages is confronted with acquiring new skills through the continuous emergence of language, perceptual-motor, cognitive and psychological functioning. This development process is shaped by the texture of the child's environment and its accompanying stress and supports.

(Phyllis, R.M., et al., 1978).

Young Children (preschoolers) :

This period extends from the end of the first year to six years of age. According to Erikson's psychosocial theory, the child becomes increasingly active, to master new skills, to be productive and acquiring a sense of initiative. In Erikson's words "initiative adds to autonomy", the quality of understanding planning and attacking the task for the sake of being on the move. The successful resolution of this stage depends on children's repeatedly attaining desired goals receiving social approval goals they have selected. The result, is children with a sense of purpose, competence and self-confidence in their ability to achieve their set goals. Repeated disapproval of self-selected goals results in a sense of defeat, guilt and resignation that prevent children from confidently selecting future goals. (Ellen, C.P., et al., 1984).

According to Piaget theory, this period is called pre-operational period in which children can think about things that are not present, their thinking is still illogical , egocentric and they still can not generalize from isolated concrete experience (Okasha, A., 1980).

According to Kohlberg theory of Moral development, this stage is called premoral stage in which child behaves appropriately because of fear of punishment or chance to receive a reward but does not recognize any higher principles. (Okasha, A., 1980).

As regards to Freud's psychoanalytical theory, this period divided into :

- (a) Oral phase : Extend from birth to 1½ years. In this phase, the child will follow the pleasure principle and immediate gratification. Firstly, he will depend on his mother's breast as the only source for his pleasure, so, if we leave the child on breast feeding for long time, it will be difficult for him to pass through further phases of development and he will always try to achieve his emotional, sexual and nutrient pleasures through his mouth i.e. oral personality.
- (b) Anal phase : Extend from 1½ to 3 years old. In this phase, the child gets his pleasure by the freedom and inability to control his anal and urinary sphinctures

If the parents try to control the child sphinctures at too early age, he will develop what is called compulsive or obessional or anal personality.

- (c) Phallic phase : Extend from 3 to 6 years. This phase starts with the ability to speak and the acquisition of the habits to satisfy his daily needs. Here the child change his source of pleasure from his anus to his genital organs and starts to be curious about differences between the man and woman. In this period, boys develop "Oedipus complex" and girls develop "Electra complex" (Okasha, A., 1980).

The School-aged child :

From six to twelve years of age, children spend an increasing proportion of their time away from their home and family, and their emotional support comes from an even-widening circle. They gain increasing independence from their parents, though still needing a secure sense of belonging. This results in strong conformity with peers and characteristic need for peer approval (Ellen, C.P., et al., 1984).

As regards to Erikson theory of psychosocial development, this stage is marked by the acquisition of sense of industry versus sense of inferiority. Children of this stage are described as believing "I am What, I learn and what

I can do". They gain recognition by being successful and their efforts and the acquisition of new skills take a primary importance. Meeting internal and external demands and criteria for performance is essential for children to negotiate successfully the choice between industry and inferiority, and to acquire a sense of themselves as adequate and competent. (Ellen, C.P., et al., 1984).

According to Piaget theory, the cognitive capabilities of school-aged children are much more sophisticated than those of preschoolers. The superficial, perceptual features of objects, people and events are no longer so central and are replaced by generalised rules and principles. These logical rules add a new stability to all their concepts including those of time and causality. Children at this stage, still continue to think very concretely, and still only about concepts and phenomena with which they have had some experience (Okasha, A., 1980).

Freud's psychosexual stage, this period latency stage, in which children learn to identify with parents of the same sex. They begin to internalize some thoughts, feelings and styles of their own sex group (Okasha, A. 1980)

In Kohlberg moral theory, this stage is called conventional conformity stage in which the child behaves as the other people's expectations for conformity to avoid disapproval and dislike by other (Okasha, A., 1980).

PSYCHOLOGICAL SEQUELAE OF CHRONIC CONDITIONS :

Chronic condition is defined as any condition persisting more than three months. (Phyllis, R.M., et al., 1978).

Another definition of chronic condition, it is a disorder with a protracted course which may be progressive and fatal or associated with relatively normal life span despite impaired physical and mental functioning. Such a disease frequently shows periods of acute exacerbation requiring intensive medical attention (Neil, K., 1979).

The effect of long-term illness and chronic condition on children and their family is pervasive. It impinges on the normal growth and development of the child; the process and quality of interrelationships and the total family-life pattern. The duration, severity and the nature of the disease are all relevant parameters in determining the psychological effects of any chronic condition, but overriding as a source of extreme stress is the uncertainty that accompanies each day (Phyllis, R.M., et al., 1978).

Prevalence of Chronic Illness :

The result of one of the few-large-scale population based studies conducted since 1975, show that among the three thousands children in the area studied, about 30 percent of children had a chronic illness. (Walker, D.K., et al., 1981). In this population, approximately 2.5 times

as many children with chronic disorders had behavioural and social problems when compared with those who were healthy. Among those with chronic conditions 8.3 percent had behavioural problems and 8.4 percent had social problems reported by their parents to trained interview. (Barry, P., 1984).

Advances in pediatric medicine have resulted in increase survival rates among children with serious life threatening disease and consequently, the numbers of youngsters continuing life with chronic debilitating illness is constantly increasing (Bedell, J.R. et al., 1977).

Many studies described frequency of psychosocial difficulties among chronic illness (Drotor, D., 1981).

There is no simple or direct linkage between physical disease and psychosocial behaviour. The relationships are multidimensional and complex (Orr, D.P., et al., 1984).

Although the effects of chronic conditions will vary significantly with the age of the child, Hughes (1976) suggested that there are eight basic emotional needs that become challenged :

- 1) Love and affection.
- 2) Security.
- 3) Acceptance as an individual.

- 4) Self respect.
- 5) Achievement.
- 6) Recognition.
- 7) Independence.
- 8) Authority and discipline.

For the child with chronic illness, fear of unknown, of pain and of death, feelings of weakness and different blocked achievement, increased dependence on parents and professionals, diminished discipline - all major threats to these needs. His development may be influenced by feelings of rejection, lack of acceptance, hostility or lack of affection, restriction in day-to-day experiences, interpersonal communication morbidity, self care, activity levels, education and plans for career and marriage are other factors that will have major effects on developmental process. (Talbot, N.B., et al., 1971).

Behaviourally, children respond to illness along the parameters of activity or passivity, with anxiety and/or depression, with withdrawal and/or anger, depending on the nature of the condition, the established personality of the child and the family's response. Stubble field, R., (1974), point, out that, inevitably, chronic illness first produce a depression reaction including shock, apathy and detachment as well as other regressive processes associated with mourning.