THE MOST COMMON ABDOMINAL TUMOURS IN CHILDHOOD

A THESIS

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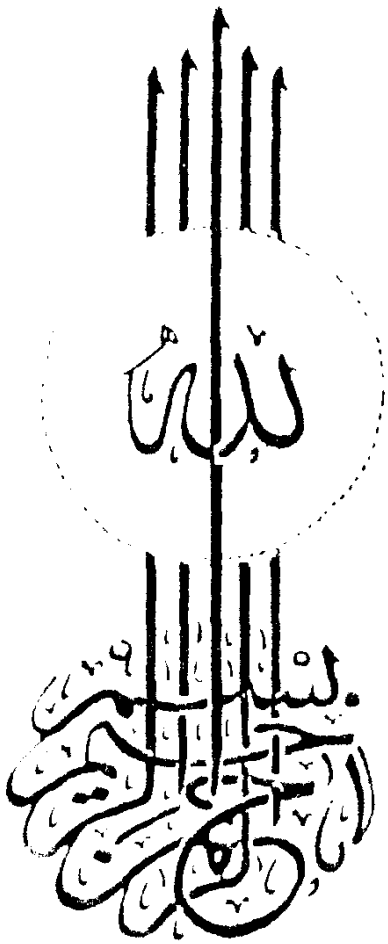

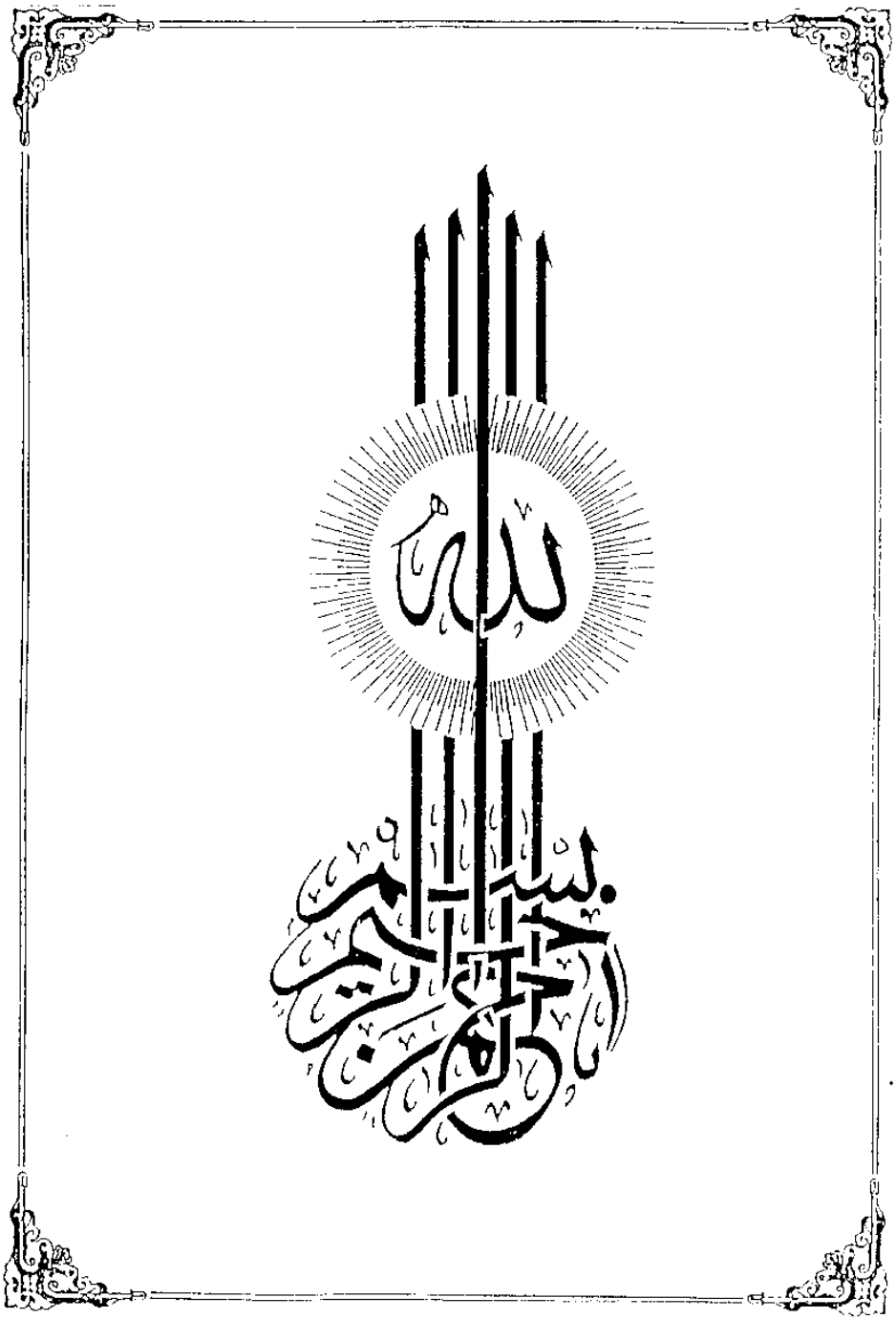

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INTRODUCTION

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Malignant disease in childhood is not so common having an approximate incidence of one in 600 children under 15 years of age or one per 10.000 children per year. In developed countries as United States and England and Wales, malignant disease is a major cause of death in children and is only exceeded by deaths from accidents.(Marsden and Steward, 1968). Acute leukaemia, lymphomas and tumours of CNS account for more than half of cancers in children. Abdominal tumours principally neuroblastoma and Wilm's tumour, constitute the next most important group (Williams & Martin, 1982).

In adults most cancers occur on such exposed surfaces as skin, lung, gut, and bladder. It is believed that most adult cancers are caused by exposure of these surfaces to a variety of environmental agents. In contrast, in children most cancers occur in deep tissues such as blood and bone marrow, brain, kidney , and skeletal tissues.(Strons,1977).

Many pediatric malignancies are of embryonal nature and a large number have their peak incidence before 5 years of age, for example neuroblastoma, Wilm's and acute lymphatic leukaemia. A proportion are associated with congenital malformations or chromosomal abnormalities and, together with an increased familial incidence in some tumours, this

suggests that prenatal factors are important in their causation. Wilm's , and primary hepatic carcinoma have been reported in the same patient having congenital malformations. (Fraumeni et al.,1968).

Knudson (1976), recently proposed a causal hypothesis for childhood cancer in which he suggests that it is dependent upon two discrete mutational events. The first mutation renders the cell precancerous, and the second transform the cell into a cancer cell: For some childhood cancers the first mutation is hereditary and only exposure to a second mutagen such as virus, radiation or chemical stimulus is required while for others the first event is a somatic mutation.

For example, diethylstilbestrol was given to mothers to prevent abortions, causing vaginal cancer in female offspring during the second decade of life. Theoretically any known human carcinogen that cross the placenta may induce cancer in the off-spring - perhaps after a latent period of several decades (Otten et al., 1977).

Early investigations indicated that there was an increased frequency of leukemia and other childhood cancer after exposure of the mother's abdomen during pregnancy to diagnostic irradiation (Bithell and Stewart, 1975). Everson (1980) suggested that cancer could result from transplacental

exposure to maternal smoking. Immunodeficiency states predispose to renal dysplasia which is associated with the development of Wilm's tumour (Penn, 1978).

No effective ways of preventing cancer are available, expect for avoiding those specific agents such as ionizing irradiation, that are known to increase the frequency of malignancy. Furthermore, the child at high risk of malignancy should have careful repeated examination in order to detect lesions at an early stage. (Strons, 1977).

AIM OF THE WORK:

To write an essay on the most common abdominal tumours in childhood. The material of the work will include the pathogenesis, clinical manifestation, laboratory data and treatment in the most common abdominal tumours as Wilm's tumour, neuroblastoma, rhabdomyosarcoma, Lymphosarcoma hepatoblastoma and teratomas.

NEUROBLASTOMA

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Neuroblastoma originates from neural crest cells which normally gives rise to adrenal medulla and the sympatheitc ganglia. (Groncy and Finkelstein, 1978)

Incidence:

Neuroblastoma is the most common malignant tumour in infancy and occurs in approximately 9.6 white children and 7 black children per 1 million in the United States each year (Young and Miller, 1975).

At diagnosis, 50 percent of the patients are under age 2, 75 percent are under age 4 and 90 percent are below 10.(Wilson and Draper, 1974).

The tumours is uncommon after 6 years of age, rare in adolescence and adulthood (Dosik et al.,1978).

Male children are slightly more frequently affected than female, in a ratio of 1.2 : 1 (Groncy & Finkelstein, 1978).

Familial neuroblastoma has been reported in several cases which suggests that rare occasions neuroblastoma may have resulted from germinal mutation. (Chatten and Voorhess, 1967).

Unlike a number of other childhood malignancies, neuroblastoma does not appear to occur more common in children with immunodeffeciency syndromes, congenital

defects, or chromosomal abnormalities. (Seeler, 1979).

It has been associated with fetal alcohol syndrome, Skull and brain defects, neurofibromatosis and Hirschsprung's disease. (Witzleben & Landy, 1974).

Robinson and McCorquodale, (1981), reported that three patients have been described with trisomy D, and neuroblastoma.

Pathogenesis:

It has been established that neuroblastoma confined to the adrenal gland can be an incidental finding at autopsy in infants less than 3 months old this finding has been termed"in situ neuroblastoma". (Beckwith & Perrin, 1963).

There is 40 fold increase in the autopsy incidence of neuroblastoma compared with the clinical incidence of the tumour, indicates that maturation of the tumour occur spontaneously in most infants, as a result of host defenses present in the infant. (Guin et al.,1969).

Pathology:

Gross Description: When the tumour is small it is well demarcated and is surrounded by a pseudocapsule. Usually the tumour is large and highly invasive with involvement of the entire adrenal gland, leaving only a narrow rim

of adrenal cortex at the periphery of the tumour. Frequently the tumour extends into the kidney, and infiltrating lymph nodes. The cut surface is usually lobulated, grey white and of a soft consistency. Areas of necrosis and calcification are frequently present (Reynold's et al.,1981).

Light microscopy:

Beckwith and Martin, (1968) have developed a histopathologic grading of neuroblastoma which demonstrate a direct correlation with survival.

Table (1) Histopathologic Grading of Neuroblastoma.

Grade	Differentiation
I	Predominantly differentiated (over 50 percent (differentiating elements)
II	Predominantly undifferentiated (5 to 50 percent differentiating elements)
III.	Slightly differentiated (less than 5 percent differentiating elements).
IV	Undifferentiated (no recognizable neurogenesis)

(Beckwith and Martin, 1968)

The diagnosis of neuroblastoma by light microscopy depends on identification of dendritic processes associated with the relatively uniform, small nuclei of neuroblasts. In the well-differentiated type, various-sized island of

round cells showing nuclear poly morphism with interspersed fibrillar areas represent aggregates of cellular processes are usually seen. Rossette formation by neuroblasts is often emphasized, but it is uncommon in the undifferentiated type. (Beckwith & Martin, 1968).

Electron microscopy: The presence of cytoplasmic processes is a constant feature that reflects the nural crest derivation of neuroblasts. The number of these processes may indicate the degree of differentiation of neuroblastoma. (Mackay et al. 1973).

Ganglioneuroblastoma:

Five to ten percent of tumours show areas of differentiation with the presence of ganglion cells, these tumours are classified as ganglioneuroblastomas. They appear in older children and adolescents. The adrenal medulla and posterior mediastinum are the most common sites of origin. (Yokayama et al.,1971).

Ganglioneuromas:

Which are usually encapsulated, represent the bengin end of the neuroblastoma spectrum. They appear more commonly in adolescents and young adults. They may produce elevation in the urinary catecholamine excretion similar to those seen with undifferentiated nueroblastomas. (Yokayama, et al.,1971).

Clinical Manifestations:

The primary tumour in the retroperitoneal region may arise in the adrenal medulla or the sympathetic ganglia, 75 percent of such tumours arise in the retroperitoneal space and over 50 percent in the adrenal gland . Many of the non specific symptoms of neuroblastoma related to abdomen, and these include anorexia, distrubances of bowel function, and abdominal pain. Palpable mass is usually the commonest presenting feature, and the mass is generally in the flank or subcostal regions. It usually crosses the midline and is firm and irregular to palpation with poorly defined margins. Abdominal distension is the presenting feature in over 50 percent of children under 1 year of age. Pain occurs due to hemorrhage into the mass. (Murthy et al.,1978).

Presacral neuroblastoma may simulate presacral teratomata, and may result in urinary frequency, inability to void due to intrinsic bladder obstruction, or constipation. (Hepler, 1976).

Symptoms due to metastasis:

Subcutaneous nodules, often with bluish color are the presenting signs in neonatal neuroblastoma, which become erythematous for 2 or 3 minutes after palpation followed by blanching as a result of vasoconstriction from the release of catecholamines from the tumour cells (Hawthorne, 1970).