

# MANAGEMENT OF PEDIATRIC RHABDOMYOSARCOMA

Essay

Submitted in Partial Fulfillment of  
Master Degree in Radiation Therapy & Nuclear Medicine

By

NIVINE MAHMOUD A. GADO  
(M.B., B. Ch.)



516.994-18  
N. M

2012/1  
15447



Supervisors

**DR. SOHEIR S. ISMAIL**

Assistant Prof. of Radiation therapy  
and Nuclear Medicine  
Faculty of Medicine, Ain Shams University

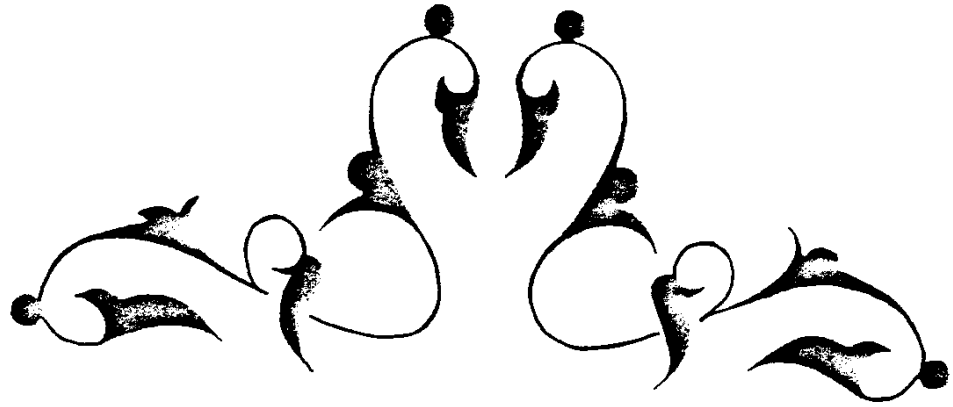
**PROF.DR.SALWA M. IBRAHIM**

Prof. of Radiation therapy  
and Nuclear Medicine  
Faculty of Medicine, Ain Shams University

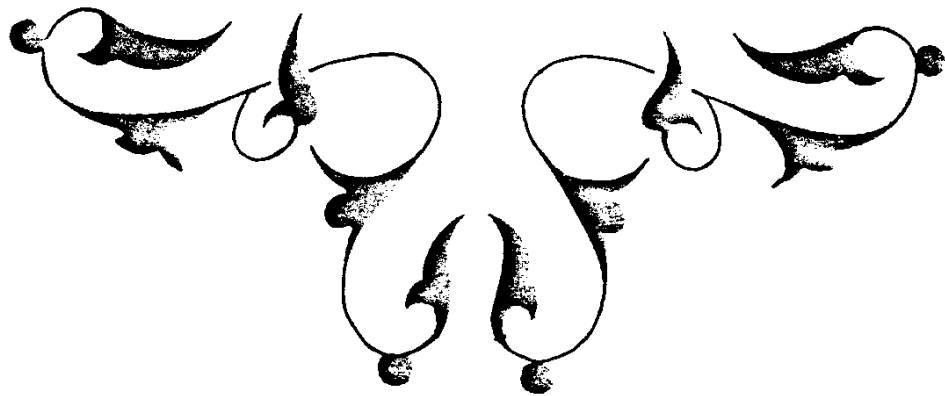
**DR. ATEF YOUSSEF RIAD**

Assistant Prof. of Radiation therapy and Nuclear Medicine  
Faculty of Medicine, Ain Shams University

Faculty of Medicine  
Ain Shams University  
1993



بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ





## ACKNOWLEDGEMENT

**I** would like to express my profound gratitude and sincere appreciation to my Prof. Dr. Laila Faris, Head of Radiation Oncology and Nuclear Medicine Department, Faculty of Medicine, Ain Shams University. I am greatly thankful for her most valuable advice, continuous encouragement and indispensable guidance.

**I** was fortunate to carry out this work under the guidance of Assistant Prof. Dr. Soheir Ismail, Assistant Professor of Radiation Oncology and Nuclear Medicine Department, Faculty of Medicine, Ain Shams University who offered me a lot of her time and experience. She contributed greatly to bring this work to its form through her suggestions, valuable observations and meticulous revision of every possible detail. To her, I owe what is beyond expression and for her no words of thanks or gratitude are sufficient.

**M**any thanks are devoted to Professor Dr. Salwa Massoud, Professor of Radiation Oncology and Nuclear Medicine Department, Faculty of Medicine, Ain Shams University for her sincere and immense help. I am greatly indebted for her valuable help during preparation of this study.

**M**y sincere appreciation goes to Assistant Professor Dr. Atef Youssef, Assistant Professor of Radiation Oncology and Nuclear Medicine Department, Faculty of Medicine, Ain Shams University, for all the help, guidance and encouragement during the preparation of the study.

**G**reat thanks are paid to the Staff members of the Department of Radiation Oncology and Nuclear Medicine and to my colleagues for their kind sympathy during the accomplishment of this work.

## LIST OF ABBREVIATIONS

|          |                                                |
|----------|------------------------------------------------|
| AFIP     | = Armed Forces Institute of Pathology.         |
| cGy      | = Centi-Grey.                                  |
| CSF      | = Cerebrospinal fluid.                         |
| CT scans | = Computerized Tomography Scans.               |
| Gy       | = Grey.                                        |
| IVP      | = Intravenous pyelography.                     |
| IRS      | = International RMS study group.               |
| Mev      | = Million electron volts.                      |
| mg       | = Milligram.                                   |
| ml       | = millilitre.                                  |
| MRI      | = Magnetic Resonance Imaging.                  |
| MV       | = Million volts.                               |
| RMS      | = Rhabdomyosarcoma.                            |
| SIOP     | = International Society of Pediatric Oncology. |
| SSD      | = Source-Skin distance.                        |
| TNM      | = Tumor-node metastasis.                       |

## CONTENTS

|                                                          |       |
|----------------------------------------------------------|-------|
| INTRODUCTION AND AIM OF THE WORK .....                   | (1)   |
| REVIEW OF LITERATURE:                                    |       |
| * Epidemiology .....                                     | (3)   |
| * Pathology .....                                        | (17)  |
| * Staging .....                                          | (34)  |
| * Clinical presentation and differential diagnosis ..... | (43)  |
| * Investigations .....                                   | (57)  |
| * Treatment .....                                        | (76)  |
| - Surgery .....                                          | (77)  |
| - Chemotherapy .....                                     | (86)  |
| - Radiotherapy .....                                     | (98)  |
| - Specific Tumor sites .....                             | (112) |
| - Therapeutic considerations .....                       | (122) |
| * Prognosis .....                                        | (126) |
| SUMMARY .....                                            | (135) |
| REFERENCES .....                                         | (139) |
| ARABIC SUMMARY.                                          |       |

# INTRODUCTION AND AIM OF THE WORK

## INTRODUCTION

Rhabdomyosarcoma is the most common pediatric soft tissue sarcoma, comprising about 50% of all soft tissue sarcomas in children, with an annual incidence of 4.5 per million in white and 1.3 per million in blacks (DeVita, 1989).

During the past decade, major therapeutic advances and, in particular, a planned multidisciplinary approach to childhood cancer have resulted in a dramatic improvement in survival and relapse-free survival rates for a spectrum of pediatric neoplasia. The long term survival rates for children with rhabdomyosarcoma have increased from approximately 10 per cent to 50 - 75 per cent range. This change reflects the widespread use of early and exhaustive diagnostic evaluation, modern surgical and radiotherapeutic techniques and effective multiagent chemotherapeutic regimens (Green et al., 1978).

As a result of these improvements, a child presenting today with newly diagnosed rhabdomyosarcoma benefits from accurate pretreatment staging, better local control of the primary tumor, and potential eradication of micrometastasis (Donaldson, 1992).

### Analysis of Childhood Cancer:

Approximately 6500 new cases of childhood cancer are diagnosed each year in the United States. Cancer is second only to accidents as the leading cause of death in children younger than 15 years of age. Leukemias and lymphomas comprise almost 48% of pediatric cancers, followed by tumors of central nervous system (20%), the sympathetic nervous system, soft tissue, kidney, bone, liver, eye and germ cells (Pizzo et al., 1989).

### AIM OF THE WORK:

The aim of this study is to review the natural history, pathology, diagnosis and results of combined modality therapy of pediatric rhabdomyosarcoma.

# REVIEW OF LITERATURE

## EPIDEMIOLOGY

### Incidence:

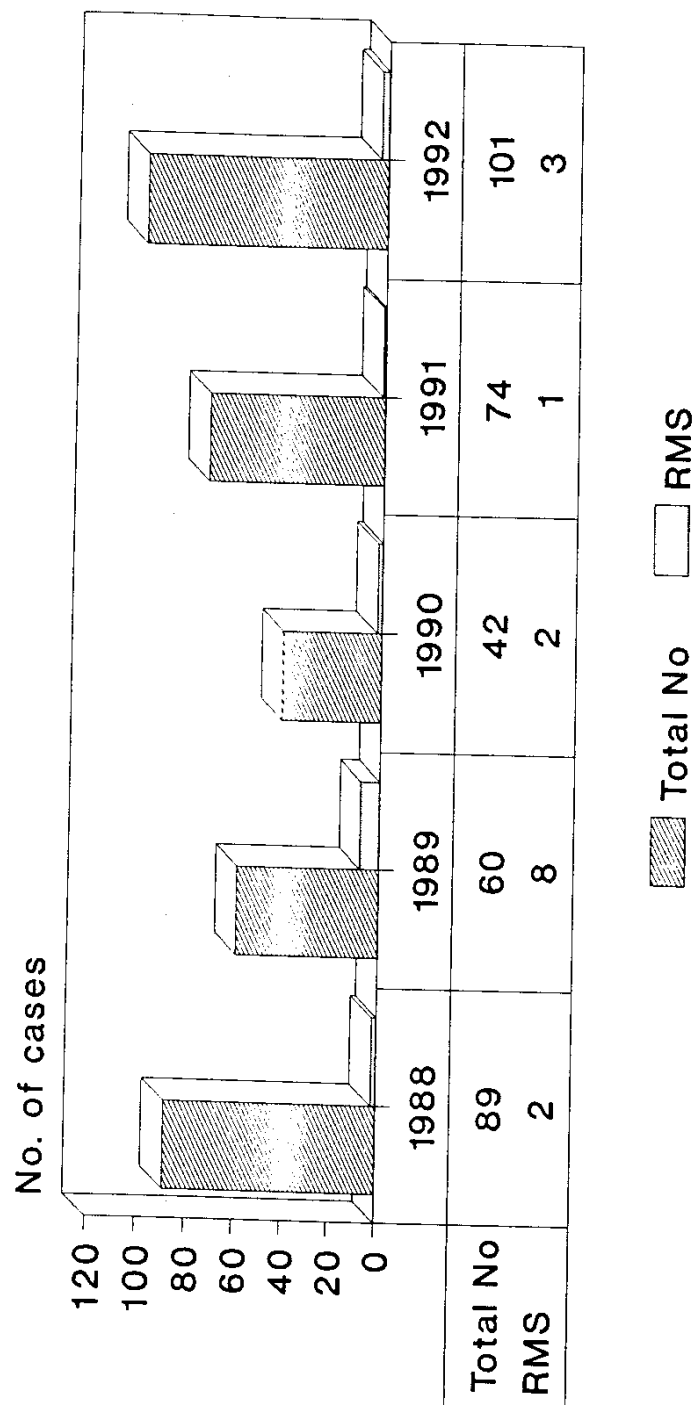
Rhabdomyosarcoma is important because of both its frequency and the multiple anatomic sites in which it can arise (Raney et al., 1989). At the United States, it is considered the sixth most common malignant tumor in children after acute leukemia, tumors of the central nervous system, lymphoma, neuroblastoma and Wilm's tumor (Hurwitz, 1990).

The annual incidence is estimated to be 4.5 per million white children and 1.3 per million black children (Pizzo et al., 1989).

It constitutes 10 - 12% of malignant solid tumors in childhood (Donaldson, 1992). It is the most common soft tissue tumor in childhood and comprises about 50% of all pediatric soft tissue sarcoma (Ruymann, 1987). Also, it accounts for 5% to 8% of all cases of childhood cancer at United States (Raney et al., 1983).

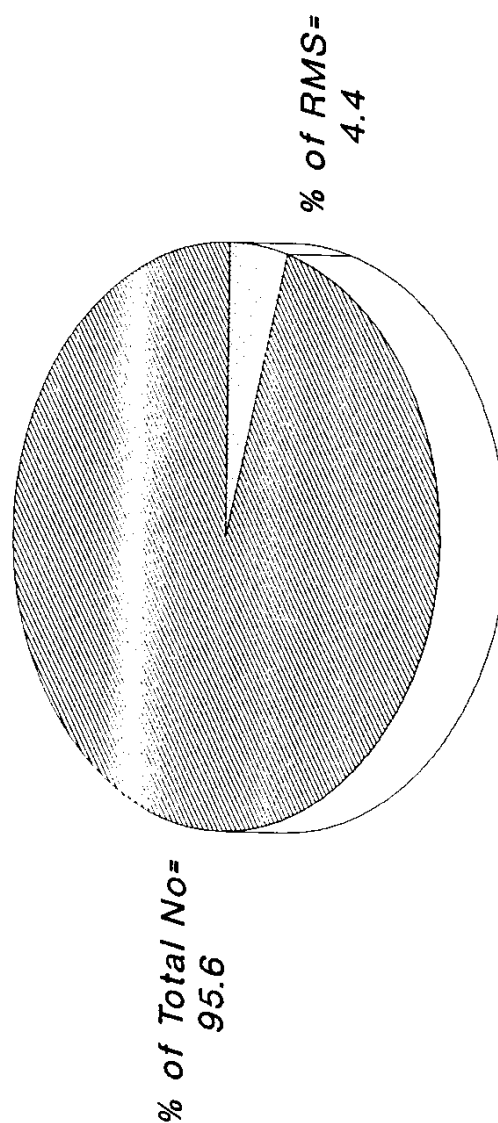
There was no significant relationship with geographic (among five continents) or change over the 20-year period (1958 - 1977) in the incidence of pediatric RMS (Breslow and Hongholy, 1983).

Fig. (1): Number of Pediatric RMS cases to Total Number of Pediatric Tumors



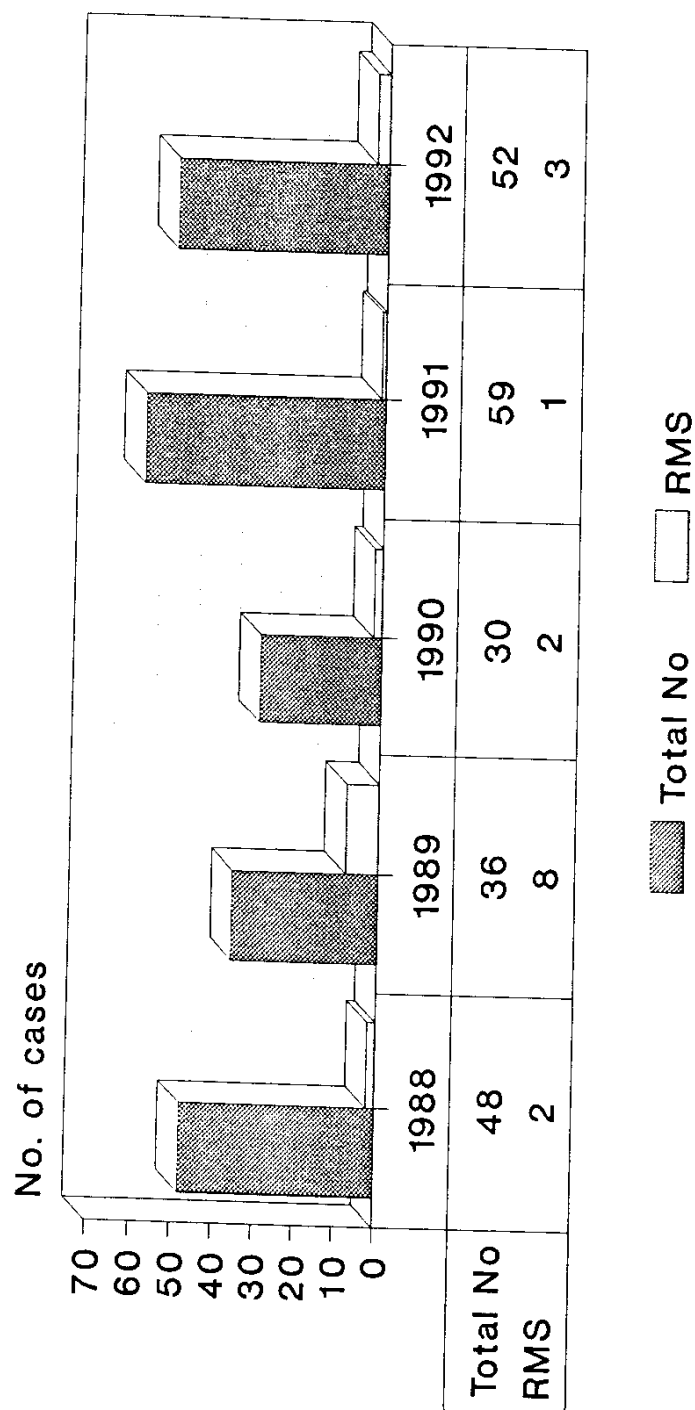
RADIO THERAPY & NUCLEAR MEDICINE DEPT.  
AIN SHAMS UNIVERSITY  
FROM 1988 - 1992

Fig. (2): Incidence of Pediatric RMS  
cases to Total Number of Pediatric  
Tumor Cases



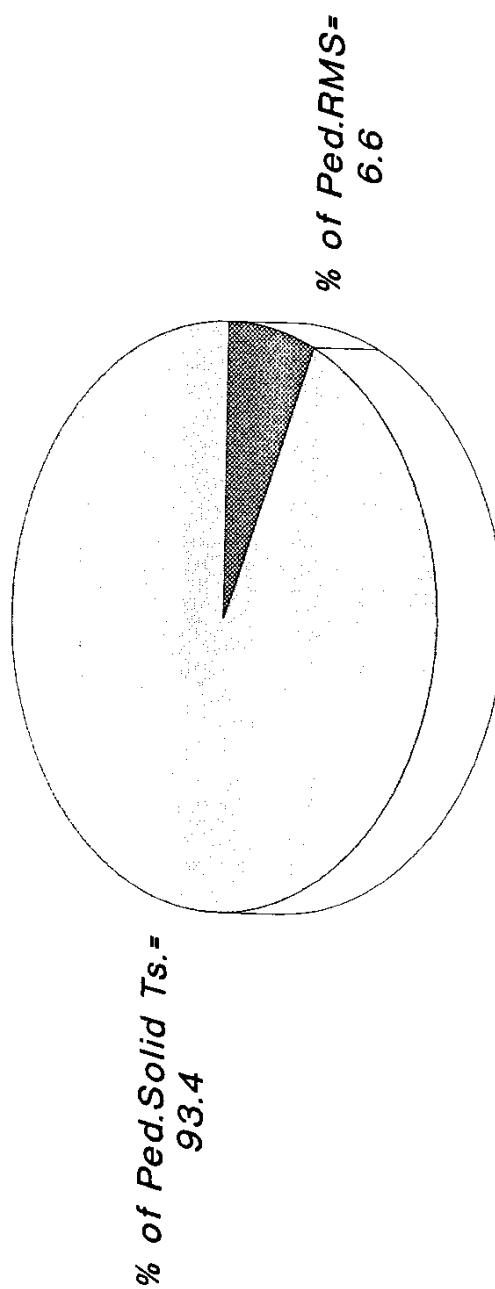
RADIO THERAPY & NUCLEAR MEDICINE DEPT.  
AIN SHAMS UNIVERSITY  
FROM 1988 - 1992

Fig. (3): Number of Pediatric RMS cases to Total Number of Pediatric Solid Tumors Cases



RADIO THERAPY & NUCLEAR MEDICINE DEPT.  
AIN SHAMS UNIVERSITY  
FROM 1988 - 1992

Fig. (4): Incidence of Pediatric RMS cases to Total Number of Pediatric Solid Tumor Cases



RADIO THERAPY & NUCLEAR MEDICINE DEPT.  
AIN SHAMS UNIVERSITY  
FROM 1988 - 1992