PEDIATRIC ORGAN TRANSPLANTATION

ESSAY

Submitted for the partial fulfilment of Master Degree in **Pediatrics**

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Dedicated

To

My Parents, And My Husband with All Affection and Respect

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Contents

Introduction	I
Aim of Essay	2
Historical Review	3
Indications of bone marrow transplantation	7
Complications of bone marrow transplantation	23
Indications of renal transplantation	33
Complications of renal transplantation	39
Indications of liver transplantation	5 0
Complications of liver transplantation	62
Indications of heart transplantation	72
Complications of heart transplantation	77
Technique of organ transplantation	82
Psychiatric aspects of organ transplantation	96
Summary	101
References	104
Arabic Summary	

Abbreviations

ABMT : Autologous bone marrow transplantation

AGL: Acute granulocytic leukemia
ALL: Acute lymphoblastic leukemia

AML : Acute myeloid leukemia Ara-C : Cytosine Arabinoside

AVNB : Avascular necrosis of bone BMT : Bone marrow transplantation

BUN : Blood urea nitrogen

CAPD : Continuous ambulatory peritoneal dialysis

CML: Chronic myeloid leukemia

CMV : Cytomegalo virus

CNS : Central nervous system

CR : Complete remission
CT : Computed tomography

Cy A : Cyclosporine A.

DMSO : Dimethylsulfoxide

DNR : Daunorubicin
EB.V : Epstein Bar virus
ECG : Electro cardiogram

EFE : Endocardial fibro-elastosis

GH : Growth hormone

GSD : Glycogen storage disease
GVHD : Graft-versus-host-disease
HLA : Human leucocytic antigen
HUS : Hemolytic uremic syndrome

ICU : Intensive care unit

ISC : Intensive sequential chemotherapy

LE: Lupus erythematosis

LPD : Lymphoproliferative disease

MHC : Major histocompatibility complexOLT : Orthotopic liver transplantation

OTC : Ornithine transcarbamylase deficiency

P.carinii: Pneumocystis carinii

PCP : Pneumocystis carinii pneumonia PEEP : Positive end expiratory pressure

PT : Prothrombin time

PTLD : Post transplantation lymphoproliferative disease

PTT : Partial thromboplastin time

SCID : Severe combined immunodeficiency

SD : Standard deviation TBI : Total body irradiation

TSH : Thyroid stimulating hormone

UW solution: University of Wisconsin solution

VOD : Veno-occlusive disease

Introduction And Aim Of Essay

Introduction

Since the start of pediatric kidney transplant program 1970, many investigators advocate children transplantation to be performed in the pediatric surgery units. (Carcassonne et al., 1989).

Recent progress in immunosuppression with cyclosporin lead the surgeon to extend the program to liver transplantation, then heart and lung transplantation (Carcassonne et al., 1989).

Extreme scarcity of small pediatric donors makes a search necessary for technical variants to benefit infants and small children from the larger group of potential adolescent and adult donors (Otte et al., 1990).

Aim of the Essay

Is to review the indications, technique, results, complications and psychologic aspects of organ transplantation in the pediatric age group.

Historical Review

Historical Review

Less than a decade ago, severe combined immunodeficiency disease (SCID) was incurable, and all children born with the disease died, usually in the first year of life (Mortimer and Alfred, 1977).

In 1968, two transplant teams reported correction of immuno-deficiency disease by successful engraftment of bone marrow from immunologically normal sibling donors (Gatti et al., 1968). Since then, transplantation of hematopoietic and lymphoid cells has been studied intensively as a treatment for SCID as well as for a large number of other diseases including acute leukemia and aplastic anemia (Mortimer and Alfred, 1977).

The history of liver transplantation dates back to the immediate post-World war II years when early experiments, which were performed primarily in dogs, showed the technical feasibility of transplanting a new functioning liver into a recipient, either heterotopically as an accessory organ or orthotopically, i.e., by complete replacement of the original organ. As might be expected, graft rejection was and still is a major problem. However, since the late 1970's, promising progress has been made in controlling this complication (Rudi, 1984).

The first human orthotopic liver transplantation was performed in 1963 by Starzl and his associates at the University of Colorado in Denver, but the first and several subsequent patients survived for only a short time. Although survival improved during the following years, the 1-year survival reported by the Starzl team in the precyclosporin era, i.e., from 1963 to the late 1970's, ranged from only 24% in adults to 33% in children. Because of these relatively discouraging results, liver transplantation during the first decade was performed almost exclusively by the Denver Center, although in 1968 Calne at the University Hospital at Cambridge and Williams at the King's College Hospital in London had set up another center which to date has performed over 130 transplantations (Rudi, 1984).

In the late 1970's, several other centers were established, among which those in Hanover and Groningen have contributed the largest number of cases. Small series of technically successful liver transplantations also have been reported from other parts of the world and most recently from other institutions in the United States (Rudi, 1984).

Recently there is a specialized center for management of liver diseases in Egypt. This center is the liver Institute - Shebeen El-Koom - Monofia University (Personal communication).

The first case of pediatric liver transplantation has been performed by Dr. Nagi Habib, an Egyptian Surgeon. It has been performed at december 1991 in the Liver Institute. The Candidate was a child 6 years old. He was suffering from glycogen storage disease complicated by liver cirrhosis. The graft has been taken from the left lobe of his mother. The operation succeeded and both the mother and the child were in a good condition post-operatively. (Personal communication).

In 1960s, transplantation of the heart, liver or pancreas was principally a laboratory event. Renal transplantation gained clinical acceptance, backed by hemodialysis as alternative renal failure therapy (Owen, 1989).

Relatively poor results in transplantation of the liver and the heart led to surgical moratoria in the 1970s. Currently, about 75% of liver transplant recipients, 75% - 80% of heart transplant recipients, and over 90% of kidney transplant recipients are alive at 1 year. (Schroeder and Hunt, 1987). Full rehabilitation occurs in two-thirds to four-fifth of those whose grafts are functioning at 1 year, as reported by clinicians at active transplant centers (Williams et al., 1987).

In the past, 10% of patients with end-stage renal disease have been treated with transplantation, but Eggers found that