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Therapeutic Cytapheresis

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Literature on therapeutic plasma exchange may be found in many different journals and books. Among the periodicals that print clinical aspects of this field, we find journals in the field of internal medicine, more especially of its subspecialties hematology, nephrology and neurology. This, of course, corresponds to the major indications for this form of treatment in patients with disorders related to the hemopoietic, kidney and nervous systems. More specialized journals directly (but not exclusively) devoted to plasma exchange are found in the fields of transfusion medicine and medicine of artificial organs.

The present issue of Current Studies in Hematology and Blood Transfusion is composed such as to allow readers an outlook into the next few years; it should help to evaluate our expectations. For comprehensive overviews on indications, the reader is referred to other books and review articles. Clinicians weigh the usefulness of therapeutic plasma exchange differently. Between 'Plasmapheresis - a treatment in search for a disease' [1] and 'plasmapheresis - formal indication in thrombotic thrombocytopenic purpura' [2], a wide span of enthusiasm can be noted. My own position in anticipation is not to withhold plasma exchange from a patient who would really benefit from it.

The major issue then, before even considering plasma exchange, is respecting the risk/benefit equation. One would think that sufficient experience for resolving this equation exists from hemodialysis but obviously patients with hyperviscosity, thrombotic thrombocytopenic purpura or Guillain-Barr disease have little in common with kidney patients. In addition, the blood volume imbalances encountered with some older but still serving plasma exchange machines are more significant than those inherent to hemodialyzers. In other words, plasma exchange has very specific side effects to consider before a patient becomes eligible. The dilemma of the risk/benefit equation is particularly stringent in the severely ill. Two extremes from our own casuistics may illustrate this:

A 23-year-old male suffering from severe autoimmune hemolytic anemia secondary to Hodgkin's disease (Hb concentration: 2.8 g/dl) was plasma exchanged using volume supplementation with red cell concentrates by the end of the procedure. He supported the treatment without side effects.
For an 80-year-old female suffering from severe drug-induced hemolytic anemia with positive indirect antiglobulin test (Hb concentration: 4.2 g/dl), plasma exchange was discussed as a therapeutic option but dismissed because of a history of cardiorespiratory insufficiency. Under protection of i.v. steroids, she received two red cell concentrates along with immunosuppressive treatment which brought her out of the acute disease phase. The 2 cases not only illustrate similar diseases managed by two different procedures (plasma exchange and red cell transfusion vs. red cell transfusion alone), but also demonstrate that not infrequently we may be called in for plasma exchange to patients who are in an acute phase of their disease. This situation may occur for two reasons:

A patient develops gradually into a critical stage. On his/her way to this stage, a number of noninvasive drug treatments are given. If they fail, plasma exchange remains as an ultima ratio procedure; wherever possible we should reject plasma exchange in these situations.

Patients with thrombotic thrombocytopenic purpura, to take this example, again, Guillain-Barr disease or hyperviscosity syndrome may be critically ill for the first time when plasma exchange is indicated, which puts the physician in a position to prescribe a relatively invasive therapy at a moment when the patients are critically ill. With modern continuous flow machines, side effects are, however, relatively rare. The advantage of the continuous flow over cyclic machines is the possibility to maintain a steady state during the entire procedure.

At the beginning of the 1990s we may also appreciate how important a treatment modality plasmapheresis has become. The initial anticipation of a frequently used therapy in a large number of diseases has emerged as an overstatement. Beyond restriction of the list of diseases eligible for treatment by plasmapheresis, we observe at the same time that those diseases that remain as formal indications are rather rare in a general hospital population. In a recent survey of the Canadian Apheresis Study Group, 55% of several hundred patients treated by plasma exchange had Guillain-Barr disease.

Preface IX

In the last 20 years, just 82 patients have been treated for thrombotic thrombocytopenic purpura in Italy [2]. If, then, the diseases we treat are rare, plasma exchange likely becomes forgotten by many physicians. This book works against it. It documents improvement of existing techniques and introduction of new ones.

By so doing, the book recalls plasmapheresis as a relative inoffensive albeit labor-intensive treatment which can help many patients on their way to improvement.
The helpful advice by Prof. A. Hassig, Bern, in the design of this book is
kindly acknowledged. The editor is indebted to Ms. U. Frauchiger for expert
editorial assistance.

U.E. Nydegger, Bern

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Abbreviations

Ab Antibody
AIDS Acquired immunodeficiency syndrome
ANA Anti-nuclear Ab
ARC AIDS-related complex
AZT Azidothymidin
CD Cluster of differentiation
CIC Circulating immune complexes
CMV Cytomegalovirus
CPA Cyclophosphamid
CR Complement receptor
CS Ciclosporin
DMS Dermatomyositis
DM Diabetes mellitus
ENA Extractable nuclear antibody
FH Ficoll-Hypaque
FNGN Focal necrotizing glomerulonephritis
FSGN Focal segmental glomerulosclerosis
GBM Glomerular basement membrane
GBS Guillain-Barr syndrome
GM-CSF Granulocyte-macrophage colony-stimulating factor
GN Glomerulonephritis
HDL High-density lipoproteins
HIV Human immunodeficiency virus
HLA Human leukocyte antigen
IFN Interferon-alpha
IL Interleukin
ITP Idiopathic thrombocytopenic purpura
IVIG Intravenous immunoglobulins
LAK Lymphokine-activated killer cells
LATS Long-acting thyroid stimulator
LCP Lymphocytapheresis
LCT Lymphocytotoxicity
LDL Low density lipoproteins
MCGN Mesangiocapillary glomerulonephritis
MCTD Mixed connective tissue disease
MEC Mixed essential cryoglobulinemia
MG Myasthenia gravis
MS Multiple sclerosis
PAF Platelet-aggregating factor
PAIg Platelet-associated immunoglobulins
PBC Primary biliary cirrhosis
PE Plasma exchange
PM Polymyositis
PN Periarteritis nodosa
PP Plasmapheresis
PSL Prednisolone
PSS Progressive systemic sclerosis
RA Rheumatoid arthritis
RANA Rheumatoid-associated nuclear antigen
RE Reticuloendothelial
RF Rheumatoid factor
RNP Ribonucleoprotein
SLE Systemic lupus erythematosus
TIL Tumor infiltrating lymphocytes
TNF Tumor necrosis factor
TTP Thrombotic thrombocytopenic purpura
VLDL Very-low-density lipoproteins
vWF von Willebrand factor
WG Wegner's granulomatosis