

# Thrombotic Thrombocytopenic Purpura: overview on the last 10 years of plasma exchange treatment in Hospital São José, Lisbon

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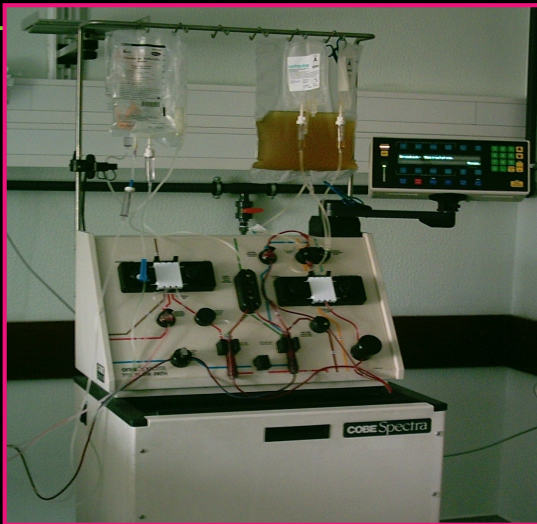


## INTRODUCTION

- Thrombotic Thrombocytopenic Purpura (TTP) was first described by Moschowitz in 1924.
- It is a rare disease.
- In the era before therapeutic plasma exchange (PE) 90% of patients died from systemic microvascular thrombosis.
- PE removes the causative antibody to von Willebrand factor cleaving metalloprotease (ADAMTS<sub>13</sub>) and replaces ADAMTS<sub>13</sub>. Recognition of TTP can be difficult because of the variety of presentations and lack of specific diagnosis criteria.
- A diagnosis of TTP may be made in the presence of a microangiopathic haemolytic anaemia and thrombocytopenia in the absence of any other identifiable cause.
- PE is the only treatment for which there are firm data on its effectiveness in TTP in adults.

## MATERIAL AND METHODS

- Patient's Hospital medical records were reviewed since January 2000 until December 2010.



- Daily PE with replacement of 1,0 to 1,5 times the predicted patient's plasma volume was initiated at least 24h after diagnosis, for a minimum of two days after platelet count and lactate deshydrogenase returned to normal.
- CobeSpectra® (Caridian) was used; the replacement fluid administered was solvent/detergent-treated plasma (Octaplas®, Octapharma); central venous access was used in all patients.

## RESULTS

- In the last ten years we performed PE in 14 adult patients (11 females and 3 males).
- 21 episodes of TTP were observed: 17 were idiopathic and 4 were related to HIV infection (1) or first trimester pregnancy (3).
- No measurements of ADAMTS<sub>13</sub> activity or antibody were made.
- Clinical presentation was heterogeneous and differential diagnosis was almost exclusively made with acute leukaemia.
- The most frequent symptom was mucocutaneous bleeding.
- Adjuvant corticosteroid therapy was instituted in all patients.
- PE was effective in all TTP episodes except one.
- Four patients relapsed.
- Four patients were submitted to immunosuppressive agents (rituximab, azathioprine and cyclophosphamide) when exacerbations or relapses occurred.
- Complications associated with PE were minor (allergic reaction and high blood pressure).

## CONCLUSIONS

- A high index of suspicion of TTP is required for rapid diagnosis and prompt initiation of PE treatment.
- PE should be instituted within 24 hours of presentation of TTP. Plasma infusion remains appropriate when there may be a delay until PE is available.
- PE is a safe and effective treatment associated with excellent clinical outcomes.
- The optimal duration of therapy is unknown and once a patient is in remission the efficacy of any treatment to prevent relapses is uncertain.