Goodpasture’s Syndrome in a 17 years girl. Fast track ICU support as bridge to full recovery after pulmonary failure.

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Abbreviations legend:
CT= computed tomography
ARDS= Acute respiratory distress syndrome
Anti GBM= anti glomerular basal membrane
C-Anca= cytoplasmatic antineutrophil cytoplasmic antibodies
P-Anca=perinuclear antineutrophil cytoplasmic antibodies
Anti PR3= against proteinasi 3
ICU= intensive care unit
CVVHDF= continuous veno-venous haemodiafiltration

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Abstract
We present a case of a 17 years girl with clinical onset of respiratory acute and severe distress, hemoptoe, plus renal failure with shock. The patient has been referred to our centre for appropriate diagnosis, critical care and medical intensive therapy, and supported to recovery successfully with a multicentric clinical assistance.

Introduction
Goodpasture’s syndrome is an uncommon and fatal disease causing haemorrage in the basement membrane lining of kidneys and lungs. By early signs and symptoms of pulmonary – renal syndrome that may lead to a diagnosis of Goodpasture’s syndrome, critical and intensive care and practice could play a key role in achieving a successful patient outcome. This very rare syndrome has an incidence of 0.5 - 1 over million people per year, with prevalence in male sex, and mainly in the 3rd and 7th decade of life. Differential diagnosis from other autoimmune diseases is necessary for stadiation of the disease and optimal therapy, and for a decision making about bioptic study.
Moreover, hemoptoe is very often an aspecifical sign in diseases such as sepsis and paraneoplastic syndrome, and is treated like an haemorragic alveolitis with more aggressive I.C.U. approach (Extra Corporeal Membrane Oxygenator), which could result non-appropriate for autoimmune diseases.

Case Report
A girl of 17 years old has been referred to the Pediatric Intensive Care Unit of our hospital with initial signs of shock, fever, low blood pressure, respiratory failure with dyspnoea, anemia, oligo-hematuria, haemoptysis, haemorragic lung relapse from airways, and requiring a complete respiratory support with mandatory mechanical ventilation, meanwhile the CT scan of chest showed a severe and bilateral alveolar shadowing (Fig. 1, 2, 3), configurating a pattern of A.R.D.S. (Acute Distress Respiratory Syndrome).
The medical history of the young patient was positive for pneumonia caused by Cytomegalovirus, (following 2 episodes of fever treated with antibiotics and paracetamol at home) requiring recovery for respiratory insufficiency and anemia.

Creatinin serum concentration was 3,4 mg/dl, Potassium 5,5 mEq/l, Haemoglobin 7 g/dl. Anti – GBM (glomerular basement membrane) selfantibodies were positive (>100 U/ml), C – ANCA (cytoplasmatic) selfantibodies positive, P – ANCA (perinuclear) negative, antiPR3 (against proteinase 3) positive (109,39 U/ml). Fibrinogen value was 398, D dimerus 10.24, configuring a pattern of intravascular disseminated coagulopathy. So, biopsy has been
deferred due the high haemorragic risk. A Goodpasture’s syndrome was diagnosed, based on clear clinical evidence, laboratories, and radiological patterns.

The patient has been treated inizialmente with full mandatory ventilatory support, and a corticosteroid therapy once antibodies presence was confirmed by lab. Ciclophosphamide 100 mg/day and azatioprine 50 mg/die was initiated. A plasma exchange with TPE 2000 SET was performed daily, associated with CVVHDF MD 100 PRE FILTRO every other day (main therapy suggested in every case of haemorragic alveolitis, even started with a Goodpasture aetiology suspected). The plasma volume exchanged daily was 2500 ml (500 ml fresh plasma + 1500 ml ringer Lactate + 500 ml Albumine 20%). At day 7, a spontaneous urine release was observed, with a serum creatinine value of 2,4 mg / dl (Table 1).

Table 1

Complete weaning from mechanical ventilation was achieved in day 8th, with complete recovery from pulmonary haemorrhage, and resolution of pulmonary shadowing at follow up X–rays of the chest.

The patient was discharged from ICU unit at day 10 without need for manteinance of dialysis, in stable chronic renal insufficiency, but with normal and spontaneous diuresis.
Pulmonary manifestations of bleeding were absent. Maintenance therapy after discharge from ICU in ward has been: prednisolon: 60 mg/die and ciclophosphamide 100 mg/die (700 mg / week) for 17 days; after chest X rays up to the present time: methyl prednison 50 mg/die and ciclophosphamide 550 mg / week plus azatioprin (150 mg /sett). At the moment, the patient is at home out of hospital, in very good clinical conditions.

Discussion
Goodpasture’s syndrome is an uncommon disorder causing haemorrhage in the basement membrane of the kidneys and lungs. Advanced care and practice nurses as well, could play a key role to a successful outcome for the patient without complications. The content of this report should stress readers about the importance of approaching the criticity of clinical onset of the disease, as it has appeared in this patient since our observation in hospital. The elapsed time from A.R.D.S. and renal failure to full recovery has been very short, and treatment with plasma exchange and medical therapy, appropriate. Furthermore, starting earlier with plasmapheresis and later with immunosuppressive agents, has shortened the recovery time of the acute phase of the disease, without risk of adverse effects. Restitution of renal function has been 100 %, but with absence of dependence from dialysis. Antibodies titre has been observing falling down by plasma exchange (Table 1), and restitution of pulmonary function to normal allowed the authors to discharge the girl from ICU to clinical follow up into clinical ward, in an interval of time of 10 days. Serum creatinin of patient is in range, and she is in reducing course of steroids. Radiographic pattern before going home, normal (Fig. 4).
Cyclophosphamide is expected to be maintained no less than 3 months. Intention of authors is to perform immunoadsorption on Protein A every 21–28 days (Fresenius Medical Care), but whether necessary³, and renal biopsy in the next future for stadiation of disease. For this reason, further communications will be available.

References