

## Vloga afereze pri akutnem sindromu Guillain-Barré

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## Akutni sindrom Guillain-Barré (GBS)

- Simetrična paraliza zaradi nenadnega vnetja perifernih živcev, ki ga povzročijo avtoimunski dejavniki kot so avtoprotitelesa
- Izmenjevalna plazmafereza
  - odstrani avtoprotitelesa in ostale možne škodljive dejavnike iz bolnikove plazme
  - je učinkovita pri bolnikih z blago, srednje hudo in hudo obliko (potrebujejo umetno ventilacijo) GBS

[Intervention review]  
**Plasma exchange for Guillain-Barré syndrome**

Jean Claude Raphael<sup>2</sup>, Sylvie Chevret<sup>3</sup>, Richard AC Hughes<sup>4</sup>, Djillali Annane<sup>1</sup>



**Authors' conclusions**

In mild Guillain-Barré syndrome, two sessions of plasma exchange are significantly superior to none. In moderate Guillain-Barré syndrome four sessions are significantly superior to two. In severe Guillain-Barré syndrome, six sessions of plasma exchange are not significantly better than four. Continuous flow plasma exchange machines may be superior to intermittent flow machines and albumin

This is a reprint of a Cochrane review, prepared and maintained by The Cochrane Collaboration and published in *The Cochrane Library* 2008, Issue 4  
<http://www.thecochranelibrary.com>

This record should be cited as: Raphael JC, Chevret S, Hughes RAC, Annane D. Plasma exchange for Guillain-Barré syndrome. *Cochrane Database of Systematic Reviews* 2002, Issue 2. Art. No.: CD001798. DOI: 10.1002/14651858.CD001798.

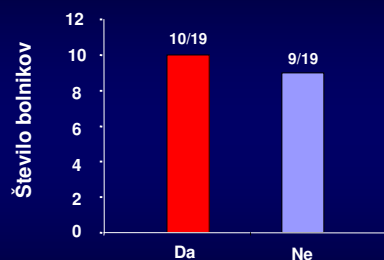
## Namen raziskave

Preučiti učinkovitost in varnost dveh afereznih metod kot sta membranska plazmafereza in imunoabsorpcija, s katerima smo zdravili bolnike s hudo obliko sindroma Guillain – Barré v času od 1998 do 2008.

## Bolniki

- 19 bolnikov s hudo obliko GBS (13 moških, 6 žensk)
- Povprečna starost: 53,7 ± 19,8 let (od 14 do 76 let)
- Vsi bolniki so imeli tetraplegijo
- 16 bolnikov je potrebovalo umetno ventilacijo

## Intravenski imunoglobulini



## Metode dela

### Membranska izmenjevalna plazmafereza (MPF)

- Plasmafilter PF 2000 N, Gambro, Švedska
- v eni proceduri se zamenja 1 to 2 volumna plazme in nadomesti z mešanico humanih albuminov in hemofiltracijsko raztopino, ki je podobna plazmini vodi (konc. albuminov je 30 g/L).

### Imunoadsorpcija (IA)

- Immunosorba®, Fresenius Medical Care, Nemčija
- 2,5 to 3 volumna plazme teče s hitrostjo 40 ml/min skozi dva imunoadsorpcijska filtra ali kolumna, ki jih sestavlja protein A

Volumen plazme izračunamo iz bolnikove višine, teže in hematokrita.

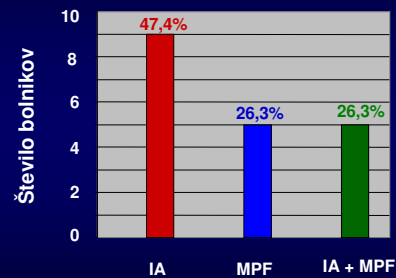
## Metode dela

- **Žilni pristop**
  - hemodializno kateter v femoralni veni pri 18 bolnikih
  - brahialna nativna AV fistula pri 1 bolniku na kronični hemodializi
- **Antikoagulacija** s heparinom
- Intenzivni hemodinamski, biokemični in klinični **nadzor** med vsako proceduro
- Odločitev glede **intenzivnosti in trajanja** afereznega zdravljenja temelji na:
  - bolnikovem kliničnem izboljšanju (potreba po umetnem predihavanju, sposobnost hoje)
  - zmanjšanju koncentracije cirkulirajočih IgG

## Rezultati

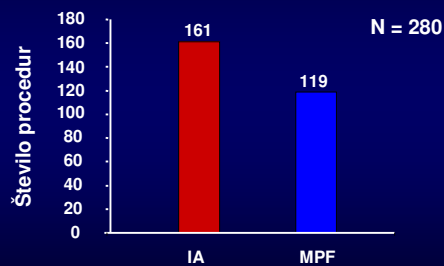
### Tip afereznega zdravljenja

(IA-imunoadsorpcija, MPF-membranska plazmafereza)



### Število afereznihih procedur

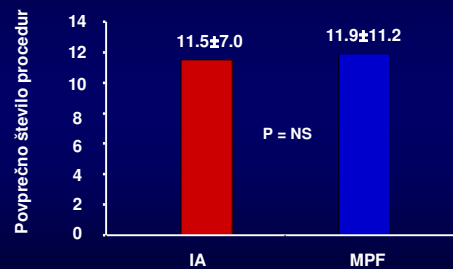
(IA-imunoadsorpcija, MPF-membranska plazmafereza)



### Povprečno število afereznihih procedur na bolnika

Bolnik s hudo obliko GBS je potreboval/prejel

4 - 30 IA in/ali 5 - 31 MPF procedur



Legenda: IA-imunoadsorpcija, MPF-membranska plazmafereza

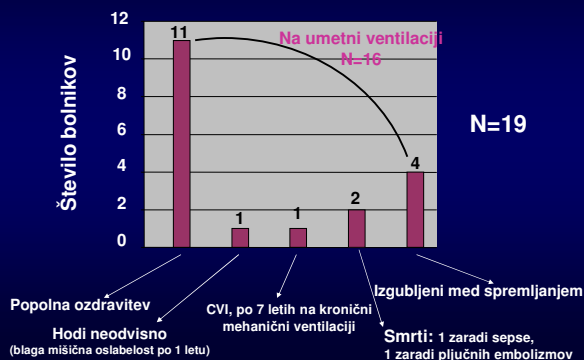
### Povprečni volumen plazme odstranjen/zamenjan med proceduro:

- 10171±1968 ml / IA proceduro (od 6500 do 13200 ml)
- 4300±1304 ml / MPF proceduro (od 3000 do 5000 ml)

### Cirkulirajoči IgG

- so se znižali z 18,9 ± 8,6 g/L na 3,5 ± 3,2 g/L (P < 0,01) po afereznem zdravljenju

### Učinkovitost afereznega zdravljenja in klinični izhod bolnikov



### Zapleti med afereznimi procedurami

- Ni bilo hude, uporne hipotenzije
- Ni bilo elektrolitnih motenj

### Zaključki

1. Pri hudi obliki GBS (potrebna umetna ventilacija) je zdravljenje z intravenskim imunoglobulinom pogosto neučinkovito. Tedaj je afereza metoda izbora.
2. Bolnike s hudo obliko GBS smo zdravili z velikim številom IA in/ali MPF procedur. Zdravljenje je bilo varno in učinkovito.
3. Naši bolniki so pogosto potrebovali in tudi bili zdravljeni z več kot arbitrarno določenimi 4 afereznimi procedurami, preden smo dosegli pomembno klinično izboljšanje.

### Bolnik z akutnim sindromom Guillain-Barre, zdravljen z dvema različnima afereznima metodama

- Ni jasno, katera aferezna metoda je bolj učinkovita za zdravljenje akutne hude oblike sindroma Guillain-Barré syndrome (GBS): plazmafereza ali imunoadsorpcija.
- Primer bolnika z rezistentno obliko GBS, ki je bil uspešno zdravljen z imunoadsorpcijo po tem, ko je bilo intenzivno dolgotrajno plazmaferezno zdravljenje neučinkovito.

### PATIENT WITH ACUTE GUILLAIN-BARRÉ SYNDROME TREATED WITH DIFFERENT APHERESIS METHODS

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#### Background

Plasma exchange therapy is the first-line treatment for severe forms of Guillain-Barré syndrome. However, it is not clear if one type of apheresis method is superior to the other. We report the case of severe Guillain-Barré syndrome, successfully treated by immunoadsorption after failure of plasma exchange therapy.

#### Methods and Results

We treated a 14-year-old male patient with acute Guillain-Barré syndrome mechanically ventilated, with two different apheresis methods. Because a course of intravenous immunoglobulin in daily dose 0.4 g/kg for 5 days didn't show any benefit, he was treated with 31 membrane plasma exchanges over 34 days. One plasma volume of 3000 ml was exchanged with diluted albumin as replacement fluid in each plasma exchange procedure. The vascular access was hemodialysis venous catheter, inserted in right and left femoral vein. Heparin was used as a standard anticoagulation method. During all sessions the patient was monitored closely and no adverse event was recorded. Sera from this patient contained a high titer of immunoglobulin G (IgG) of 32 g/L before plasmapheresis treatment, which was reduced to 0.32 g/L after 31 sessions of plasma exchange. However, his neurological deficit was still severe and he needed mechanical ventilation. Since the plasma exchange treatment was not effective, he was then switched to immunoadsorption (IA) apheresis method. A plasma volume processed was 9,000 ml per IA treatment. He received 7 sessions of IA in 15 days. After the seventh IA he didn't require assisted ventilation anymore and was able to stand up without assistance. After two years he recovered from disability completely.

#### Conclusion

The effect of plasmapheresis was incomplete in our Guillain-Barré syndrome patient suggesting that immunoadsorption may be the first line therapy for those who do not respond to the plasma exchange treatment.

- 14-letni bolnik z akutnim hudim GBS: tetraplegija, umetna ventilacija

• Zdravljenje:

1. intravenski imunoglobulini 0,4 g/kg/dan 5 dni
2. Membranska plazmafereza:  
31 procedur v 34 dneh  
3000 ml plazme zamenjan z raztopino albuminov  
(2x fem. kat., heparin)  
IgG: 32 g/L pred MPF → 0,32 g/L po MPF



Nevrološka prizadetost še vedno huda, potrebna umetna ventilacije

3. Imunoadsorpcija:  
7 procedur v 15 dneh  
9000 ml plazme prečiščene med vsako proceduro



Umetna ventilacija ni več potrebna, vstane brez pomoči  
Po 2 letih popolna ozdravitev

## Zaključek

Pri hudi obliki akutnega GBS čimprej intenzivno zdravljenje s plazmaferezo do kliničnega izboljšanja.

Če tega ne dosežemo, je smiselno poskusiti z imunoadsorpcijo.

Peritoneal Apheresis and Dialysis 13(4):310-313  
doi:10.1111/j.1744-0077.2009.00730.x  
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### Guillain-Barré Syndrome Treated by Membrane Plasma Exchange and/or Immunoadsorption

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**Abstract:** In this report, we evaluate the success of immuno-adsorption and plasma membrane exchange treatment in patients with severe Guillain-Barré syndrome (GBS), and describe one case successfully treated by immuno-adsorption after failure of plasma exchange therapy. Nineteen severely paralyzed GBS patients, aged 14-76 years, who were treated between 1995 and 2008, were retrospectively studied. They underwent 161 immuno-adsorption and 119 plasma exchange procedures. In severe GBS, 4-30 immuno-adsorption and 5-33 plasma exchange sessions were needed. Among 16 patients on mechanical ventilation, 11 recovered from disability completely, while in one patient mild muscle weakness persisted after one year. One patient suffered from ischemic stroke, two patients died after weaning from mechanical ventilation. One death was believed to be procedure-unrelated, the other patient died from sepsis. Four patients were lost to follow-up. We

treated a 14-year-old, mechanically-ventilated patient using two different apheresis methods. After failure of 31 membrane plasma exchanges over 34 days, the patient was then switched to the immuno-adsorption apheresis method, receiving seven sessions in 15 days. He no longer required assisted ventilation and recovered from the disability completely. A high number of immuno-adsorption as well as membrane plasma exchange treatments can be safely and effectively applied in severe GBS patients. Our patients often needed, and were provided with, more than the currently arbitrarily accepted four apheresis sessions before good clinical recovery was achieved. For non-responders to plasma exchange treatment, immuno-adsorption may be the first-line therapy. **Key Words:** Guillain-Barré syndrome, Intensive care, Intravenous immunoglobulin, Mechanical ventilators, Plasma exchange.