**Supplement material**

**Table legends**

**Table S1:** Difference of propensity score-matched variables and other variable between group with and without plasma exchange therapy

PE, plasma exchange; SCr, serum creatinine

**Table S2:** Case reports or series for IgAN patients with plasma exchange therapy

\*Patient also treated with eculizumab after plasma exchange; SCr ; serum creatinine; PE, plasma exchange; HSN, henoch-schonlein nephritis; eGFR: estimated Glomerular Filtration Rate.

**Figure legends**

**Figure S1:** Plasma active complement products level in patients dependent and independent of dialysis at the last follow-up

**Figure S2:** Complement activation products in patients with crescentic IgAN and other IgAN

 IgAN, IgA nephropathy; PE, plasma exchange; SCr, serum creatinine

**Table S1: Difference of propensity score-matched variables and other variable between group with and without plasma exchange therapy.**

|  |  |  |  |
| --- | --- | --- | --- |
|  | **PE group(n=12)** | **Control group(n=12)** | **P** |
| **PS matching variables** |  |  |  |
| **SCr (**μmol/l**)** mean (SD) | 705.3 (296.4) | 661.7 (249.4) | 0.71 |
| **Pulse methylprednisolone** | 9 | 9 | 0.99 |
| **Steroids + immunosuppression** | 9 | 9 | 0.99 |
| **Other variables** |  |  |  |
| **Age(ys)** mean (SD) | 42.7 (15.0) | 40.8 (15.5) | 0.77 |
| **Sex (male)** | 9 | 11 | 0.21 |
| **MABP (mmHg)** mean (SD) | 111(13) | 111(14) | 0.97 |
| **Hypertension** | 11 | 12 | 0.99 |
| **RPGN/ARF** | 11 | 11 | 0.99 |
| **Oliguria** | 2 | 2 | 0.99 |
| **Proteinuria (g/d)** median, (IQR) | 5.8 (4.5-8.7) | 4.7 (4.0-5.8) | 0.41 |
| **Alb (g/l)** mean (SD) | 26.6 (3.3) | 29.6 (4.5) | 0.14 |
| **Dialysis at presentation** | 8 | 9 | 0.65 |
| **Total crescent (%)** | 64.4 (24.4) | 69.5 (25.5) | 0.63 |
| **Tubular atrophy/interstitial fibrosis > 50% (N, %)** | 6(50%) | 8(66.7%) | 0.41 |

PE, plasma exchange; SCr, serum creatinine

**Table S2: Case reports or series for IgAN patients with plasma exchange therapy**

|  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Study** | **No.** | **Age/Gender** | **Initial renal function** | **Renal biopsy** | **Pulse of steriod** | **Oral Prednisone** | **Immunosuppression** | **Initial PE from Kidney biopsy or after admission(d)** | **Courses of PE** | **Duration of PE treatment** | **Mean replacement fluid liters/course**  | **Follow-up** | **Renal outcome** |
| **cases of crescentic IgAN** |
| Coppo, 1985 | 1 | 18/F | (CrC 13 ml/min, proteinuria 3-6 g/day | IgAN 2/12 Glomeruli hyalinized,10/12 glomeruli crescents | Yes | Yes | Yes | 7 | 18 | 10 weeks | 1 patient's plasma volume | 10 months | Decrease of IgA immune complexes; with complete recovery of renal function  |
| 2 | 54/M | CrC 15 ml/min, proteinuria 6 g/day | IgAN, 30 of 50 glomeruli with numerous fibrous crescents, remaining glomeruli showed active lesions.  | No | Yes | Yes | 7 | 9 | 4 weeks | 1 patient's plasma volume | 5 months | CrC and IgA immune complexes did not have significant change during PE. Entered ESKD after 5 months. |
|  | 3 | 21/F | SCr 425 μmol/L, proteinuria 4.6g/d | Focal segmental mesangial proliferative IgAN,5/15 Glomeruli sclerotic, 10/15 Glomeruli crescents | Yes | Yes | Yes | 21 | 6 | 2 weeks | 3L | 12 months | Renal function stabilized for 9 months, then progressed to ESKD |
| Rafael, 1990 | 4 | 54/M | SCr 796 μmol/L | IgAN 60% crescents, severe mesangial proliferation without arterial or tubular lesion | No | Yes | Yes | NA | 20 | 2 weeks | 2-3L | 6 months | After first cycle of PE, renal function recovered and without crescents and traces of IgA and C3 by biopsy at the 1st month of follow-up; No response to the second cycle of PE |
| Roccatello, 1995 | 5 | 16/M | SCr 884μmol/L | IgAN with 90% florid crescents, 10% fibrotic crescents | Yes | Yes | Yes | NA | 22 | NA | 1 patient's plasma volume | 36 months | ESKD |
| 6 | 61/F | SCr 636μmol/L | IgAN with 70% florid crescents,5% glomerular hyalinosis | Yes | Yes | Yes | NA | 14 | NA | 1 patient's plasma volume | 12 months | ESKD |
| 7 | 39/M | SCr 238μmol/L | IgAN with 50% florid crescents,35% glomerular hyalinosis | Yes | Yes | Yes | NA | 15 | NA | 1 patient's plasma volume | 12 months | ESKD |
| 8 | 55/M | SCr 654μmol/L | IgAN with 40% florid crescents,35% glomerular hyalinosis | Yes | Yes | Yes | NA | 10 | NA | 1 patient's plasma volume | 36 months | Stable SCr, moderate renal impairment |
| 9 | 18/F | SCr 265μmol/L | IgAN with 80% florid crescents,5% glomerular hyalinosis | Yes | Yes | Yes | NA | 23 | NA | 1 patient's plasma volume | 120 months | Stable SCr, moderate renal impairment |
| Yamazki, 1995 | 10 | 7/M | CrC 7.6ml/min | IgAN with 70% crescents of 43 glomeruli.  | Yes | Yes | Yes | 16 | 6 | 2 weeks | NA | 2 months | Renal function recovered to the normal range, second biopsy demonstrated a marked decrease in histological activity . |
| Chambers, 1999 | 11 | 18/M | CrC<10ml/min, SCr 2033μmol/L, proteinuria 5g/d | 31 glomeruli, 80% crescents, diffuse mesangial proliferation IgAN, focal global sclerosis, marked interstitialfibrosis, severe tubular atrophy | No | Yes | Yes | NA | 7 | 16 | 4L | 2 months | No improvement, ESKD |
| Fujinaga, 2007 | 12 | 5/M | SCr385μmol/L | IgAN with (80%) cellular crescents， fibrouscrescents, glomerulosclerosis, and interstitial fibrosis  | No | Yes | No | 13 | 5 | 10 days | 50 ml/kg | 3 months | Within normal limits chemistry findings，second biopsy with focalsegmental mesangial proliferative IgAN nephropathy, and 55%small fibrocellular crescents  |
| Shenoy, 2007 | 13 | 13.5/F | eGFR 28ml/min.1.73m2 | HSN 53% crescents | No | No | No | NA | NA | 2 weeks | 90 ml/kg  | 2.6 years | Referred promptlywere had a normal eGFR at 113 (range 98–142) ml/min per 1.73 m2 |
| 14 | 8.4/F | eGFR 64ml/min.1.73m2 | HSN 52% crescents | No | No | No | NA | NA | 2 weeks | 90 ml/kg  | 5.5 years |
| Ring 2015\* | 15 | 16/M | SCr 98µmol/L,albumin of 19 g/L | HSN with epithelial crescents in 6 of 14glomeruli | Yes | Yes | Yes | NA | 5 | NA | 40 mL/kg | about 13 months | after eculizumab treatment, renal function improved remarkably, second biopsy after 11 months revealed chronicity renal function slowly declined to end-stage at the second year  |
| **cases of non-crescentic IgAN** |
| Nicolls, 1985 | 1 | 22/M | SCr 1.89mmol/L | Primary IgAN,13/15 Glomeruli sclerotic,remaining 50% (1/2) with cellular crescent | No | Yes | Yes | NA | NA | 2 months | NA | 2 months | Rapid progression to ESKD |
| 2 | 25/M | SCr 0.84mmol/L | Primary IgAN, 25/28 Glomeruli sclerotic, 2/28 Glomeruli crescentic, 11/28 Glomerulus advanced segmental scarring | No | Yes | Yes | NA | NA | 1 month | NA | 10.5 months | Renal function stabilized for 2 mo; progressed to ESKD in 10 months |
| 3 | 16/M | SCr 0.23mmol/L | Primary focal segmental proliferative IgAN; 14/31 Glomeruli sclerotic, 8/31 Glomeruli crescents | No | No | Yes | NA | NA | 2 months | NA | 13 months | Renal function stabilized for 3 mo, then progressed to ESKD |
| Coppo, 1985 | 4 | 54/M | CrC 19 ml/min, proteinuria 4-6 giday | IgAN, 6 of 30 (20%) glomeruli with florid mostly circumferential epithelial crescents | No | Yes | Yes | 9 | 12 | 7 weeks | 1 patient's plasma volume | 12 months | Decrease of IgA immune complexes; with complete recovery of renal function  |
| 5 | 55/M | CrC 37 ml/min | Did not have another biopsy after CrC slowly and progressively fell ml/min  | No | Yes | Yes | NA | 12 | 8 weeks | 1 patient's plasma volume | 10 months | CrC and IgA immune complex reversed by PE, but returned to the previous values shortly after discontinuation.  |
| 6 | 52/M | CrC 28 ml/min, proteinuria 10g/day | Did not have another biopsy after CrC slowly and progressively fell ml/min  | No | Yes | Yes | NA | 9 | 5 weeks | 1 patient's plasma volume | 11 months | Unchanged IgA immune complex throughout the PE, Proteinuria remained in the nephrotic range and the CrC decreased further in the follow-up. |
| Lai,1987 | 7 | 24/F | SCr 727 μmol/L | IgAN, 13/20 Glomeruli sclerotic,7/20 Glomeruli with fibrocellular crescents | No | Yes | Yes | 10 | 20 | 8 month | 3L | 14 months | Renal function stabilized with PE for 10 months; progressed to ESKD in 1 year |
| Nicholls, 1990 | 8-20 | 26(17-58)/10 M | Mean SCr340(160-650)μmol/L | IgAN Median 34%(7-80%) crescents in nonsclerosed glomeruli (median, 34%; range, 7-80%). | No | No | Yes | NA | 21 | 3 months | 3-4L | 39(5-54) months | Rate of renal functional deterioration was slower than during the pre-PE period in 10 patients, four patients remain off dialysis, dialysis can be delayed. |
| Tsunoda, 1995 | 21 | 9/F | 64.4(31.3)ml/min/1.73m2, mean proteinuria 3.75(1.8)g/d | IgAN with mean crescents 23.5(5.23)% | No | No | No | Mean 4.6m | 10~16 | NA | 50-100ml/kg | >2 years | ESKD |
| 22 | 11/M | No | No | No | CRF |
| 23 | 16/M | Yes | Yes | Yes | non-CRF |
| 24 | 8/M | No | No | No | non-CRF |
| 25 | 15/M | No | Yes | Yes | non-CRF |
| Afessa, 1997 | 26 | 66/M | SCr 486μmol/L | Segmental necrotizing IgAN and crescent formation | Yes | Yes | No | 5 | 11 | 4 weeks | NA | 13 months | Stable SCr of 2.2 to 2.5mg/dL |
| Chambers, 1999 | 27 | 27/M | SCr 248μmol/l, proteinuria 6.2g/d | IgAN with 10 glomeruli, 3 cellular crescents,moderate interstitial fibrosis and tubularatrophy  | Yes | Yes | Yes | NA | 6 | 18 | 4L | 2 weeks after PE | Entered ESKD after 2 weeks  |
| Roccatello, 1995 | 28 | 44/M | SCr 106μmol/L | 40% florid crescents,15% glomerular hyalinosis | Yes | Yes | Yes |  | 11 | NA | 1 patient's plasma volume | 24 months | Slight elevation of SCr from 106 to 132μmol/L |
| Tsunoda, 1995 | 29 | 16/M | No | 64.4(31.3)ml/min/1.73m2, mean proteinuria 3.75(1.8)g/d | No | No | No | Mean 4.6m | 10~16 | NA | 50-100ml/kg | >2 years | ESKD |
| 30 | 10/M | No | No | No | No | ESKD |
| 31 | 9/F | Yes | No | No | No | ESKD |
| 32 | 12/F | No | No | No | No | ESKD |
| 33 | 14/F | No | No | No | No | ESKD |
| 34 | 8/M | No | No | No | No | ESKD |
| 35 | 18/M | No | No | Yes | No | ESKD |
| 36 | 11/M | Yes | No | No | No | CRF |
| 37 | 10/M | No | No | Yes | No | non-CRF |
| 38 | 16/M | NA | Yes | Yes | CTX | non-CRF |
| 39 | 11/M | No | No | No | No | non-CRF |
| 40 | 8/M | Yes | No | No | No | non-CRF |
| 41 | 17/M | NA | No | Yes | CTX | non-CRF |
| 42 | 15/M | Yes | No | Yes | CTX | non-CRF |
| 43 | 16/M | No | No | Yes | No | non-CRF |
| 44 | 10/M | No | No | Yes | Aza  | non-CRF |
| 45 | 13/M | No | No | Yes | Aza  | non-CRF |
| Shenoy, 2007 | 46 | 11/F | eGFR 46ml/min.1.73m2 | HSN 20% crescents | No | No | No | 6 (range 2–13) days afterreferral.  | 17 (9–28)  | 2 weeks | 90 ml/kg  | 7.5 years | One late-referred child needed renal transplant, 15 children referred promptlywere had a normal eGFR at 113 (range 98–142) ml/min per 1.73 m2, normal or only a slightlyelevated UA/UC |
| 47 | 6.8/F | eGFR 82ml/min.1.73m2 | HSN 40% crescents | No | No | No | 1.1 years |
| 48 | 5.8/M | eGFR 93ml/min.1.73m2 | HSN 24% crescents | No | No | No | 2.1 years |
| 49 | 15/M | eGFR 20ml/min.1.73m2 | HSN 20% crescents | No | No | No | 2.5 years |
| 50 | 3.7/F | eGFR 136ml/min.1.73m2 | HSN 0% crescents | No | No | No | 6.2 years |
| 51 | 12.5/F | eGFR 61ml/min.1.73m2 | HSN 43% crescents | No | No | No | 3.1 years |
| 52 | 11.8/M | eGFR 33ml/min.1.73m2 | HSN 0% crescents | No | No | No | 3.8 years |
| 53 | 12.3/M | eGFR 90ml/min.1.73m2 | IgAN 10% crescents | No | No | No | 1.1 years |
| 54 | 10.1/F | eGFR 42ml/min.1.73m2 | IgAN 29% crescents | No | No | No | 4.2 years |
| 55 | 13.1/M | eGFR 17ml/min.1.73m2 | HSN 5% crescents | No | No | No | 3.4 years |
| 56 | 9.9/M | eGFR 43ml/min.1.73m2 | HSN 14% crescents | No | No | No | 5.2 years |
| 57 | 8.3/F | eGFR 22ml/min.1.73m2 | HSN 0% crescents | No | No | No | 4.3 years |
| 58 | 8.9/F | eGFR 67ml/min.1.73m2 | HSN 0% crescents | No | No | No | 5.4 years |
| 59 | 7.7/F | eGFR 29ml/min.1.73m2 | HSN 26% crescents | No | No | No | 6.3 years |
| Augusto, 2011 | 60 | 51.9(25.4)/4M | SCr 1.06 (0.71-3.9)mg/dl proteinuria 4.3 (2.1-7.2) | Diffuse endocapillary proliferation [class 3] alone or with extracapillary proliferation [class 4] | 7 patients | Yes | No | 8 days | 12 | 66 days | mean 3.2L | 6(1-21.6) years | 2 died, 1ESKD, BVAS improved, median eGFR and proteinuria were83(22) mL/min/1.73 m2 and 0.14(0.1) g/d |

IgAN, IgA nephropathy; \*Patient also treated with eculizumab after plasma exchange; SCr ; serum creatinine; PE, plasma exchange; HSN, henoch-schonlein nephritis; eGFR: estimated Glomerular Filtration Rate; .CrC, creatinine clearance; CRF, chronic renal failure; ESKD, end stage kidney disease.

**Figure S1 Plasma active complement products level of patients dependent and independent of dialysis at the last follow-up.**





**Figure S2. Active complement components in patients with crescentic IgAN and non-crescentic IgAN**

