Supplemental data

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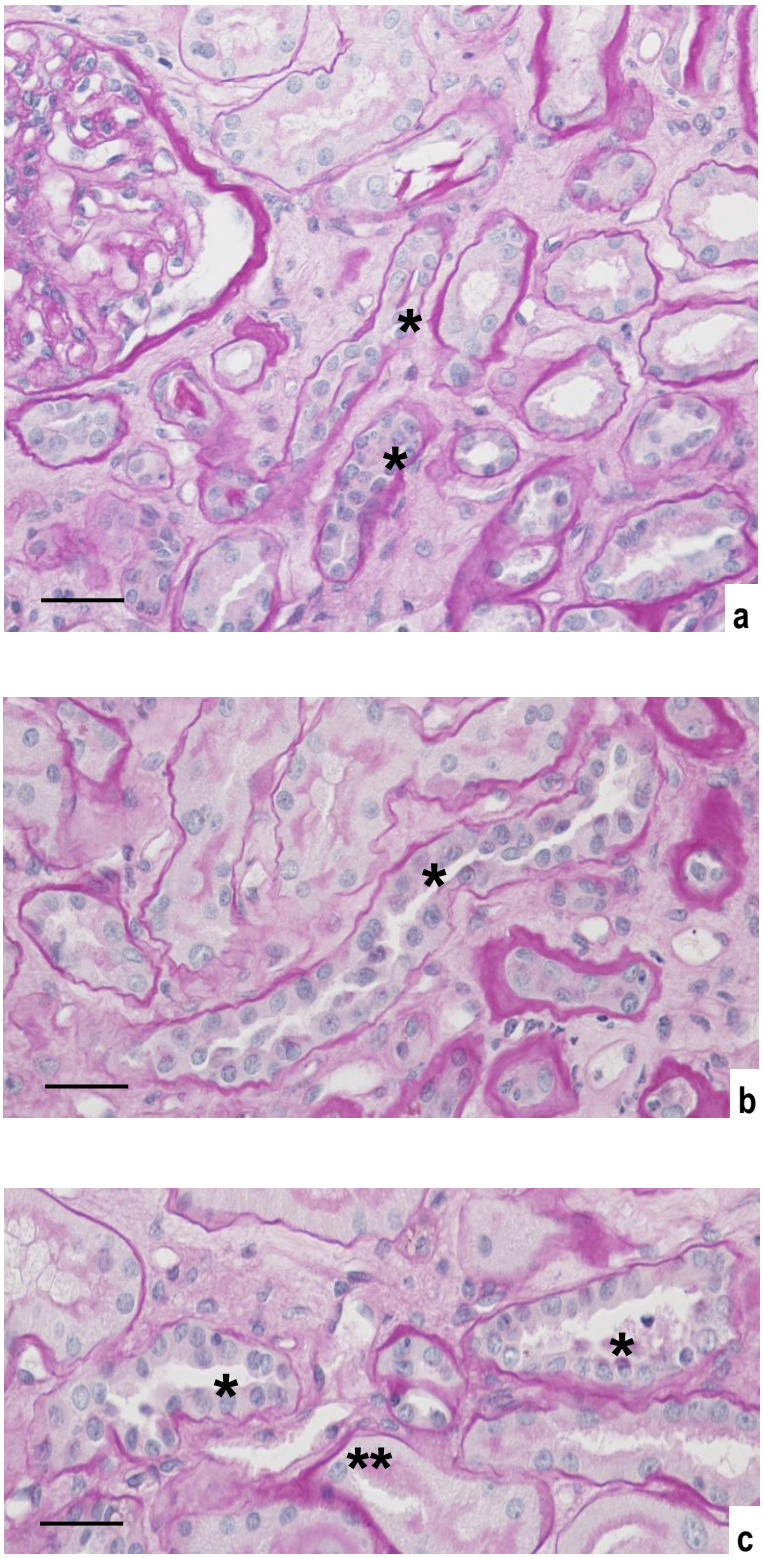
A



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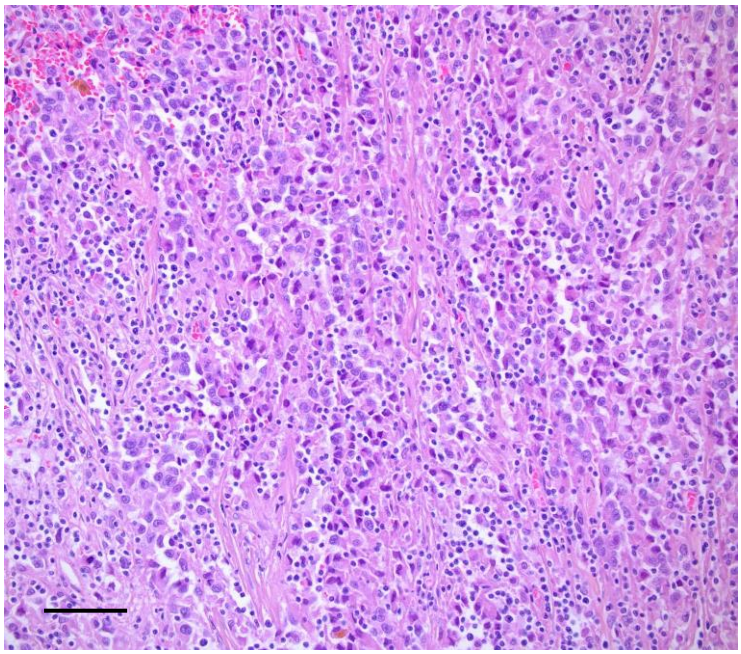
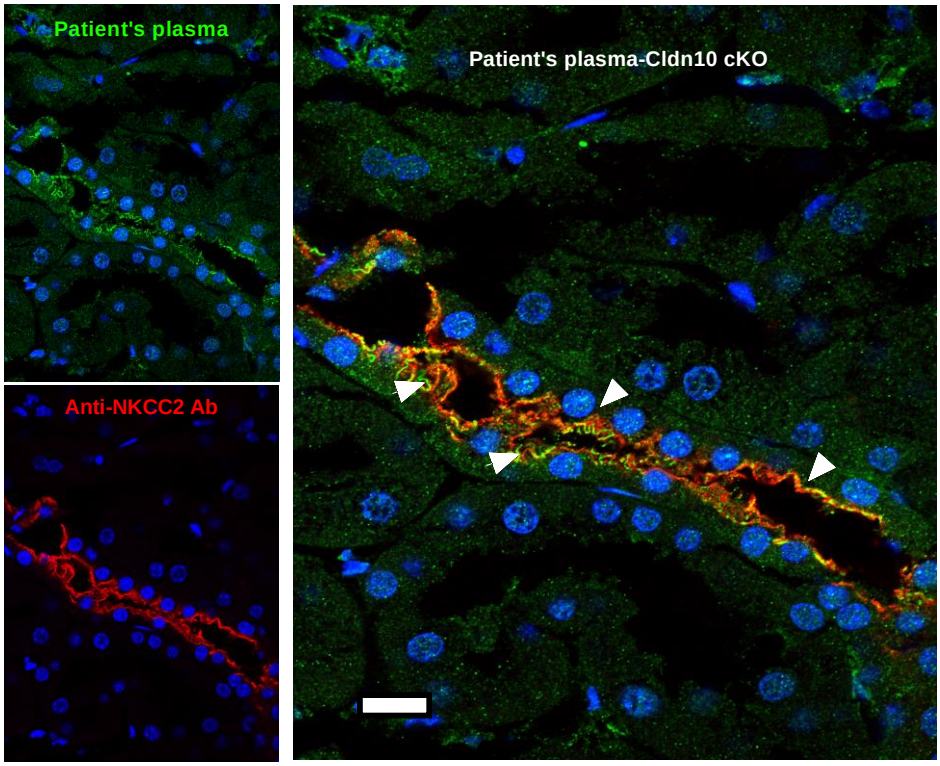


Supplemental Figure S1

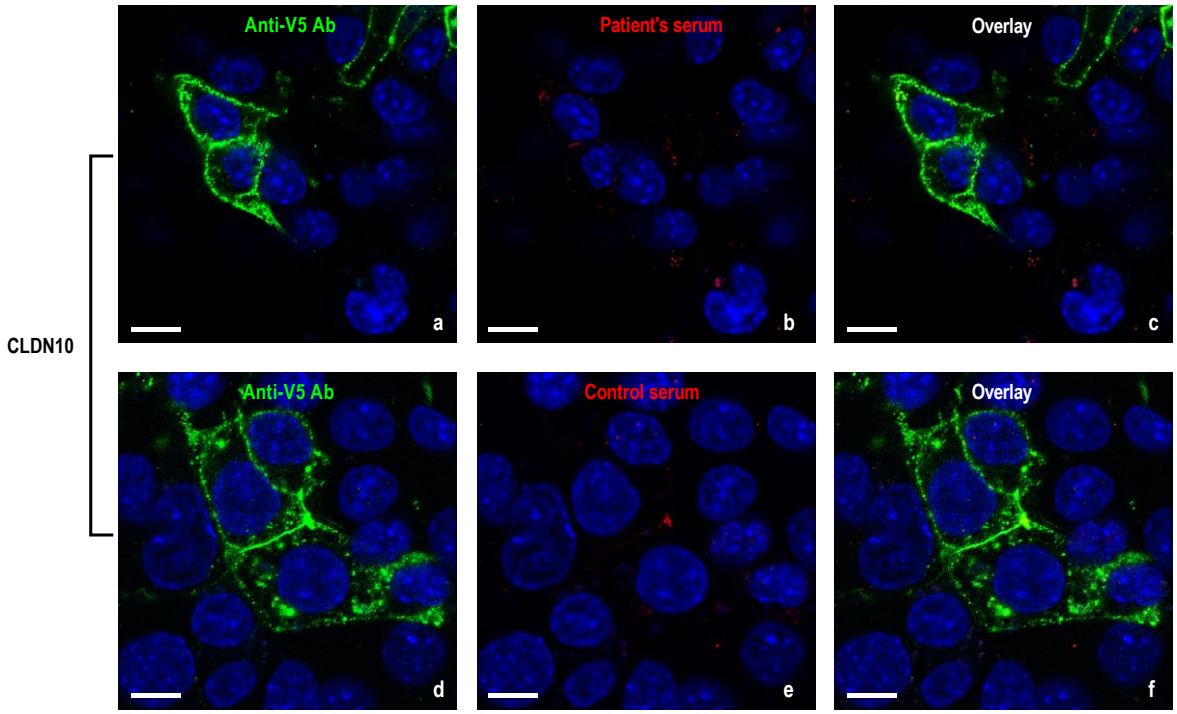
A. A kidney biopsy performed 8 months after the patient's initial presentation (light microscopy study, periodic acid-Schiff stain. Original magnification: a x100 (Bar = 100 µm), b and c x200 (Bar = 200 µm.). Features of acute tubular injury and nuclear dystrophy (*) were present in non-proximal tubules along with peritubular capillaries dilatation (**) and moderate (30-40%) interstitial fibrosis without any significant interstitial inflammatory cell infiltrate. Glomeruli (n=22, 4 sclerotic) had a normal appearance and moderate arteriosclerosis was present.

B. Piece of right nephrectomy (light microscopy, hematoxylin and eosin stain, x200). Presence of an undifferentiated grade 4 renal cell carcinoma with rhabdoid features (10%) and necrosis (10%). (Bar = 200 µm.).



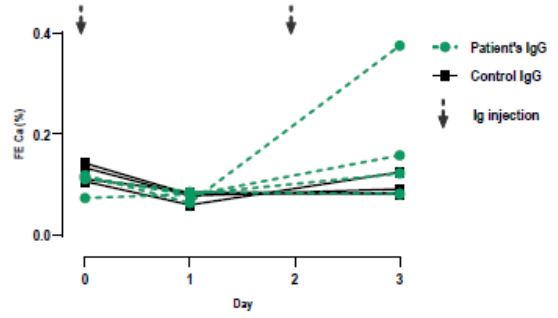
Supplemental Figure S2

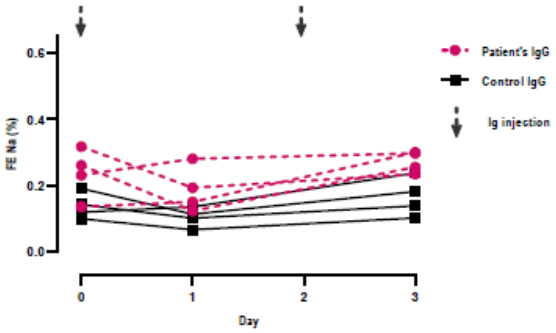
Sections of fixed fresh frozen kidney from claudin-10 conditional knock-out (Cldn10 cKO) mice were incubated with the patient's plasma (1:500), and rabbit anti NKCC2 antibody (1:7500) and subsequently with Alexa 555-conjugated goat anti-human IgG (1:600) and Alexa 488-conjugated goat anti rabbit (1:500) antibodies. Patient's plasma stained junctional domain of tubular sections expressing NKCC2 in Cldn10 cKO mice kidney (arrowheads). Bar = 20 µm.

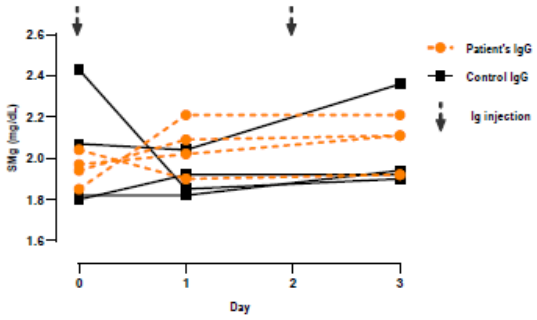


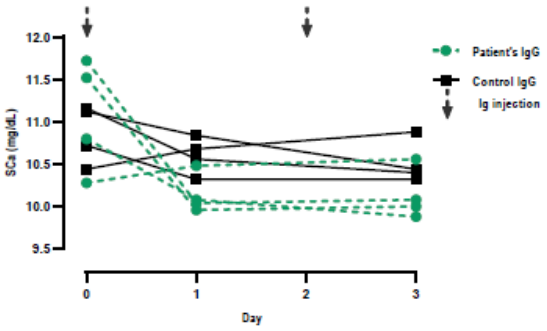
Supplemental Figure S3

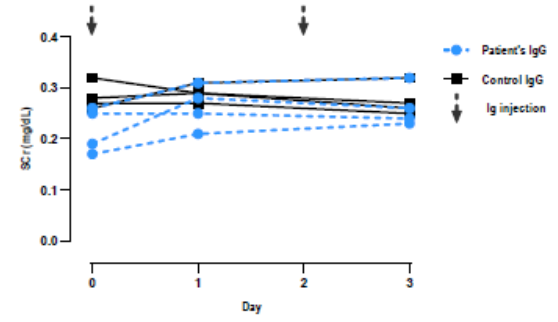
Transfected immortalized mouse kidney TAL (MKTAL) cells expressing V5-tagged human claudin 10 (CLDN10) (stained with anti-V5 Ab) (a, d) were incubated with either patient's plasma (b) or control plasma (e). Patient's plasma and control plasma did not stain plasma membrane expression CLDN10 (overlays, c, f). Bar = 10 µm.

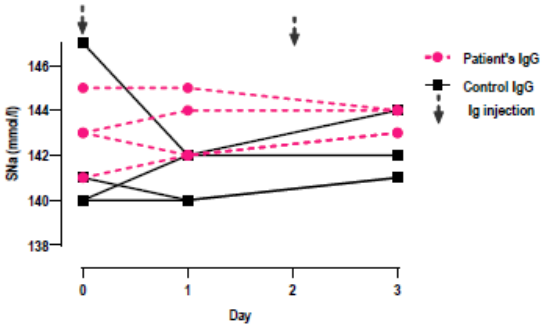

FE Ca (%)
0.4
0.2
0.0
0
1
2
3
Day
Patient's IgG
Control IgG
Ig injection


FE Na (%)
0.6
0.4
0.2
0.0
0
1
2
3
Day
Patient's IgG
Control IgG
Ig injection


SMg (mg/dL)
2.6
2.4
2.2
2.0
1.8
1.6
0
1
2
3
Day
Patient's IgG
Control IgG
Ig injection

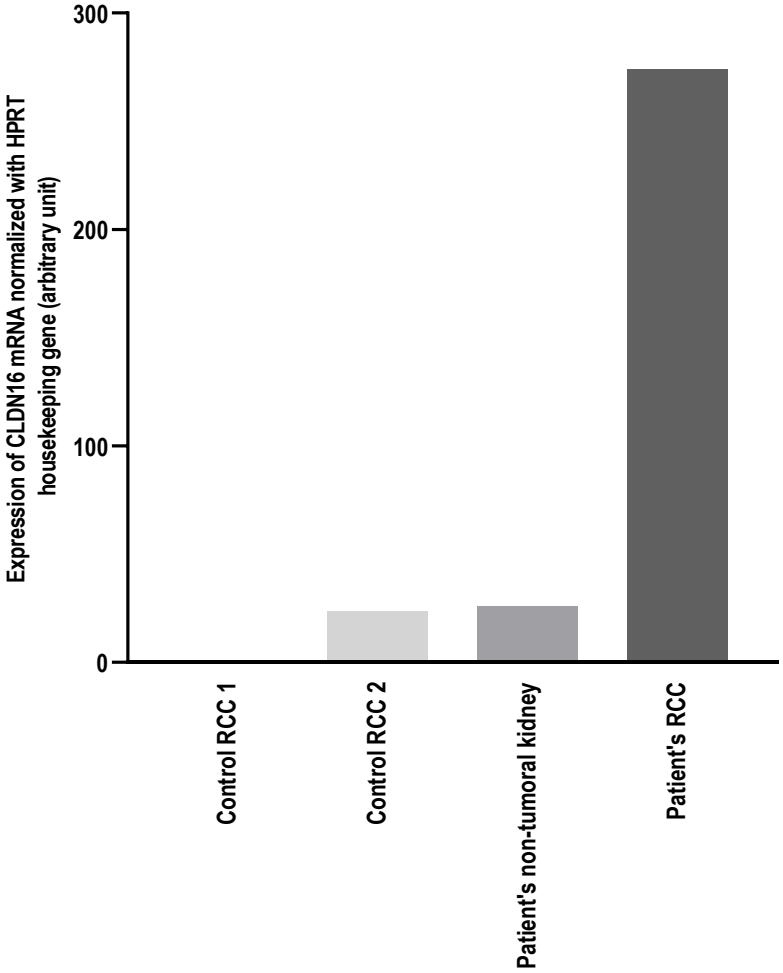

SCa (mg/dL)
12.0
11.5
11.0
10.5
10.0
9.5
0
1
2
3
Day
Patient's IgG
Control IgG
Ig injection


SCr (mg/dL)
0.4
0.3
0.2
0.1
0.0
0
1
2
3
Day
Patient's IgG
Control IgG
Ig injection


SNa (mmol/l)
146
144
142
140
138
0
1
2
3
Day
Patient's IgG
Control IgG
Ig injection

Supplemental Figure S4

IgG purified from the patient's serum and from the serum of three patients with chronic tubulo-interstitial nephropathy of undetermined cause were injected intravenously in male Sprague-Dawley rats at day 0 and day 2 (n=4, in each group). Renal fractional excretion of calcium (FECa) and sodium (FENa) and serum magnesium (SMg), calcium (SCa), creatinine (SCr) and sodium (SNa) levels were monitored throughout day 0 to day 3. None of these parameters changed throughout the experiment



Supplemental Figure S5

Claudin-16 coding mRNA was amplified by Taqman real-time PCR (Applied Biosystems, Foster City, CA, USA) in the patient's renal cell carcinoma (RCC) and non-tumoral renal tissue, and in two control RCC.