Supplemental data

Table of Contents

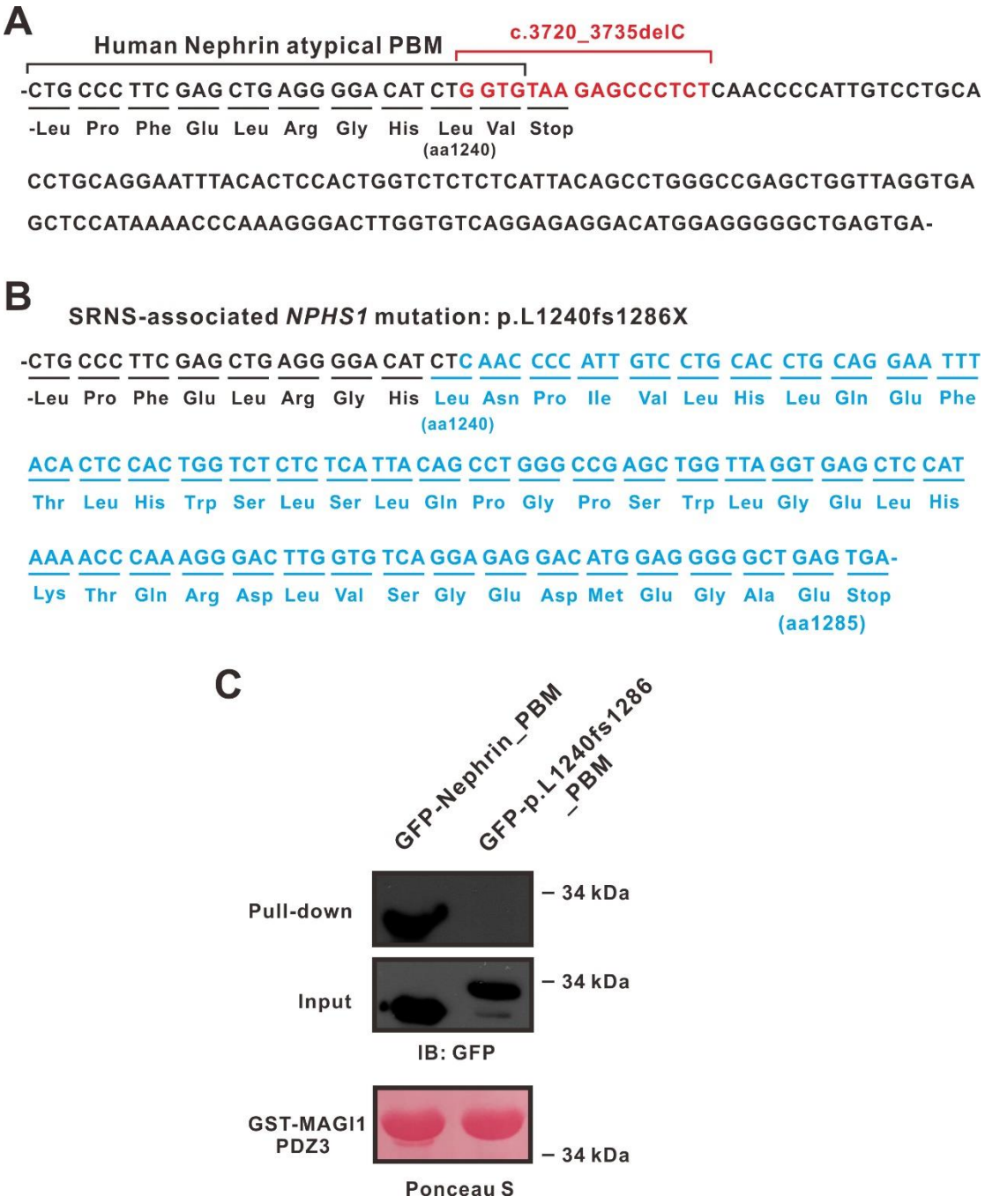

Figure S1. The SRNS-associated *NPHS1* mutation p.L1240fs1286X impairs PDZ-binding motif (PBM) that is required for MAGI1 binding. (A-B) Amino acid sequence analysis of the SRNS-associated *NPHS1* mutation p.L1240fs1286X. The 16 deleted nucleotides (c.3720_3735C) in the disease mutation are shown in red (A). The additional coding sequence and corresponding amino acids are shown in cyan (B). (C) GST-pull down assay showing that the PBM of the p.L1240fs1286X mutation could not bind to MAGI1-PDZ3.

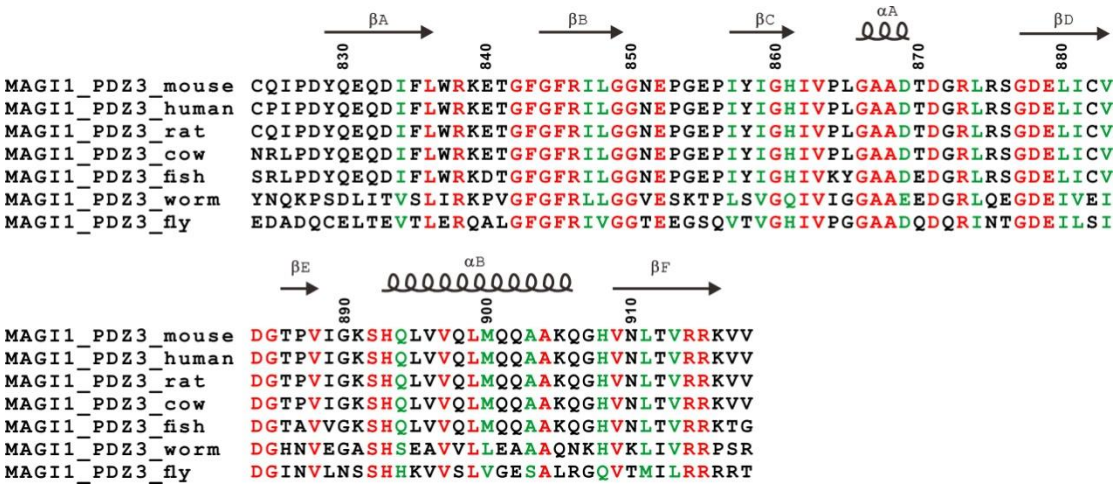

Figure S2. Amino acid sequence alignment of PDZ3 domain of MAGI1 from different species. The totally conserved and conserved residues are color in red and green, respectively.

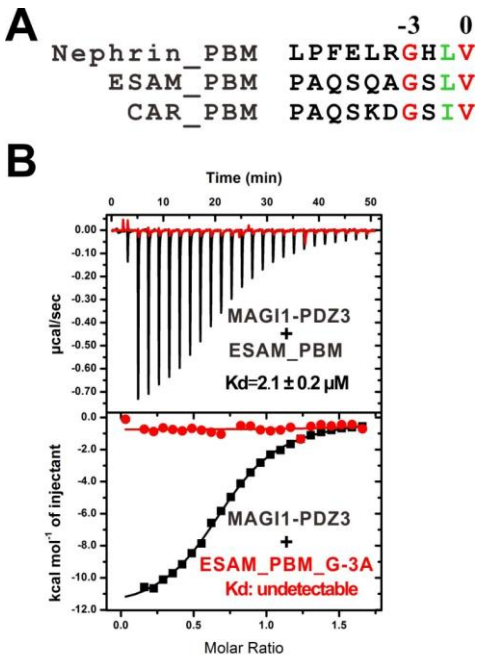

Figure S3. Gly at -3 position of PBM is the key determinant for other targets recognition by MAGI1-PDZ3. (A) Sequence alignment of PBMs of other targets of MAGI1 PDZ3. (B) ITC-based affinity measurements of bindings of MAGI1-PDZ3 to wild type and G-3A mutant of ESAM-PBM.

Table S1. Data collection and refinement statistics

Data collection and processing	
Complex	Nephrin-PBM/MAGI1-PDZ3
Source	SSRF-BL18U1
Wavelength(Å)	0.97791
Space group	I23
Unit cell(a,b,c,Å)	93.731, 93.731,93.731
Unit cell(α,β,γ,。)	90.000,90.000,90.000
Resolution range (Å)	50.00–1.78 (1.81-1.78)
No. of unique reflections	13086 (683)
Redundancy	3.9 (3.6)
I/σ(I)	16.16(5.44)
Completeness (%)	98.1 (99.9)
R_{merge} (%)[a]	6.9 (17.2)
Wilson_B	18.68
Structure refinement	
Resolution (Å)	46.865-1.780 (1.917-1.780)
R_{work}[b]/R_{free}[c] (%)	17.41 (17.44)/19.85 (21.24)
rmsd bonds (Å)/angles (°)	0.009/1.369
Number of reflections	
Working set	13083(2490)
Test set	639 (129)
Number of protein atoms	856
Number of solvent atoms	88
Average B factor (Å^2)	22.9
Ramachandran plot(%)	
Most favored regions	98.96
Additionally allowed	1.04
Generously allowed	0

Numbers in parentheses represent the value for the highest resolution shell.

[a]$R_{merge} = \sum|I_i - I_m|/\sum I_i$, where I_i is the intensity of the measured reflection and I_m is the mean intensity of all symmetry related reflections.

[b]$R_{cryst} = \Sigma||F_{obs}| - |F_{calc}||/\Sigma|F_{obs}|$, where F_{obs} and F_{calc} are observed and calculated structure factors.

[c]$R_{free} = \Sigma T||F_{obs}| - |F_{calc}||/\Sigma T|F_{obs}|$, where T is a test data set of about 5% of the total reflections randomly chosen and set aside prior to refinement.