Supplementary Table 1. Pearson correlations for selected independent variables.					
Parameter	Age (yr)	Height (cm)	MMA_S (μmol/L)	MMA_U (mmol/mol Cr)	Creatinine (mg/dL)
Height (cm)	0.8548 (<0.0001)				
MMA_S (μmol/L)	0.2348 (0.0373)	0.1937 (0.0959)			
MMA_U (mmol/mol Cr)	-0.2555 (0.0230)	-0.3062 (0.0043)	0.2791 (0.0153)		
Creatinine (mg/dL)	0.5026 (<0.0001)	0.4723 (<0.0001)	0.8075 (<0.0001)	0.0389 (0.7338)	
Cystatin C (mg/L)	0.0812 (0.6535)	0.0681 (0.7066)	0.7684 (<0.0001)	0.0096 (0.9583)	0.7901 (<0.0001)

Supplementary Table 1. Pearson correlations between age, height, serum methylmalonic acid (MMA_S), urine methylmalonic acid (MMA_U), serum creatinine (Cr) and cystatin C. *p*-values in (). Many values such as height and age (0.8231) demonstrate multicollinearity.

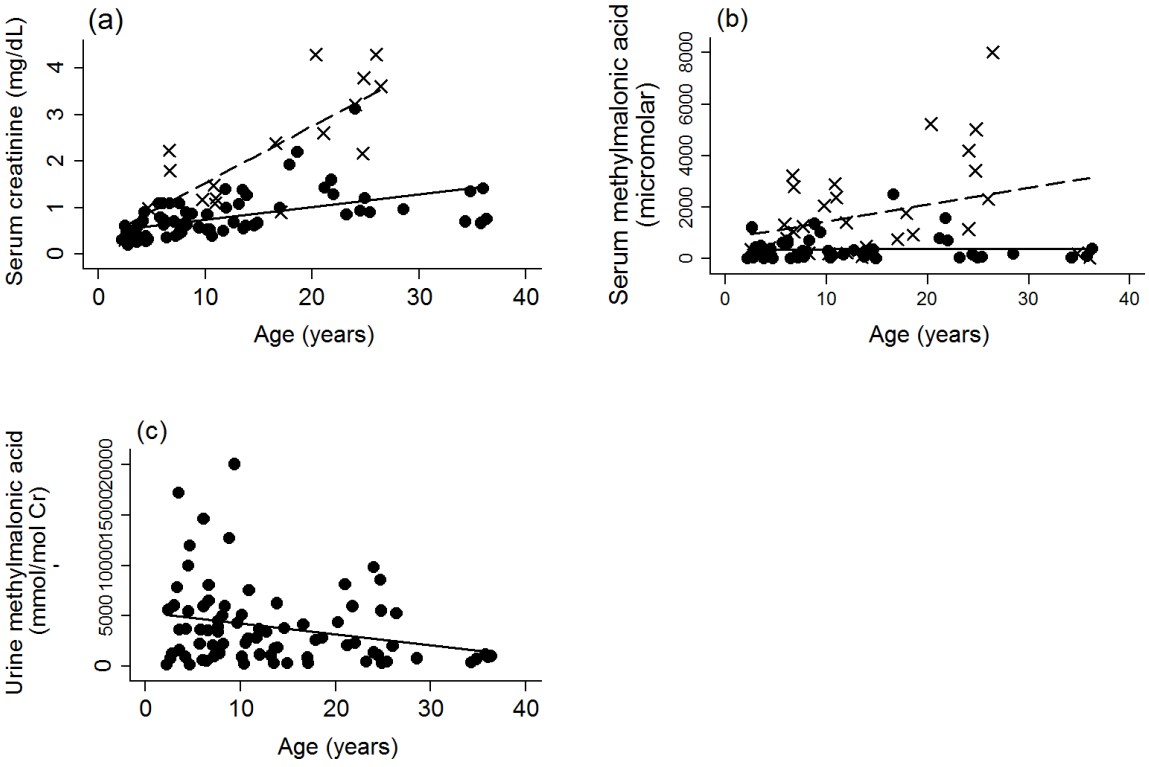
Supplementary Table 2. Correlation Coefficients				
Serum Creatinine	Correlation Coefficient (R^2)			
(mg/dL)	MMA_S, Ht	Serum cystatin C, Ht	Serum creatinine, Ht	eGFR creatinine - cystatin C, Ht
< 1.2	0.737	*	0.762	*
< 1.5	0.727	0.723	0.750	0.656
< 2.0	0.733	0.745	0.755	0.680
< 2.5	0.733	0.766	0.757	0.720
all values	0.703	0.767	0.714	0.713

Supplementary Table 2. Comparison of correlation coefficients (R^2) for regressions using models based on serum methylmalonic acid (MMA_S), serum cystatin C, serum creatinine, and eGFR creatinine - cystatin C stratified by creatinine levels. Insignificant models (*Beta* coefficients with $p > 0.05$) marked with an *.

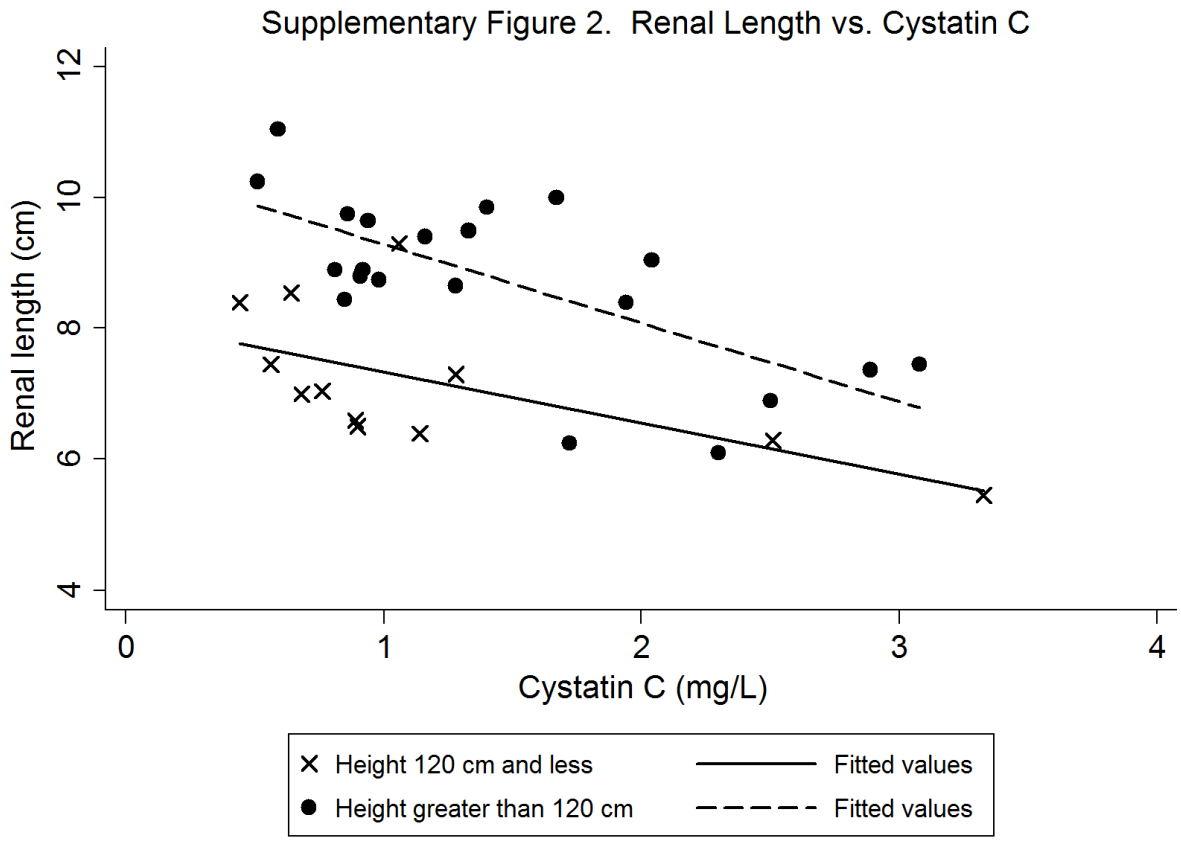
Supplementary Table 3.				
Enzymatic subtype	Observations	Serum Creatinine	Serum Methylmalonic acid	Urine Methylmalonic acid
mut				
≤ 10 years	34	0.71 (0.4-0.9)	661.3 (214-820)	5414 (2041-6000)
11 to 20 years	17	1.08 (0.62-1.28)	855.9 (221-1198)	2852.8 (1441-3739)
>20 years	12	2.49 (1.29-3.71)	2628 (663-4610)	4380 (1224-5716)
cblA				
≤ 10 years	4	0.308 (0.26-0.36)	15.0 (12.3-20.6	661.3 (365-903)
11 to 20 years	5	0.818 (0.6-0.9)	51.6 (31.1-72)	1495.4 (289-3294)
>20 years	5	0.98 (0.7-1.36)	157.1 (46-214)	1185.2 (731-973)
cblB				
≤ 10 years	2	0.64 (0.41-0.87)	1273.5	12660
11 to 20 years	1	1.93	1786	2585
>20 years	4	1.25 (1.03-1.37)	373.5 (42-755)	1268 (413.5-2140)

Supplementary Table 3. Means and interquartile range (in parenthesis) of serum and urine laboratory values stratified by enzymatic subtype and age. Serum creatinine units are given in mg/dL (reference values are age/gender dependent); serum methylmalonic acid in umol/L (reference: ≤0.40 umol/L); urine methylmalonic acid in mmol/mol creatinine (reference: <3.60 mmol/mol creatinine).

Supplementary Figure 1. Laboratory measurements vs. Age



Supplementary Figure 1. (a) Serum creatinine vs. age. Circles and solid fitted line represent patients with serum methylmalonic acid levels below 2000 micromolar and X's and dashed lines represent serum methylmalonic levels above 2000 micromolar; (b) Serum methylmalonic acid vs. age. Circles and solid fitted line represent patients with eGFR creatinine levels above 60 mL/min/1.73m^2 and X's and dashed lines represent eGFR creatinine below 60; (c) Urine methylmalonic acid vs. age.



Supplementary Figure 2. Renal length (average of left and right), stratified by height, is inversely proportional to cystatin C. This graph represents the most predictive renal length model, incorporating the predictors cystatin C and height.