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Now let's focus on classic Fabry males. When the kidney disease, it start? Well, it starts in the newborn. We have newborns, or we know newborns who already have glycolipid deposits in podocytes. We know that in childhood, at age five, six, seven, there is already evidence of podocyte injury. So there is foot process effacement in some podocytes Some podocytes have already lost their ability to prevent urinary plasma proteins, such as albumin, from leaking into UT.

So naturally history of Fabry nephropathy, first you have pathological histology without any outside evidence of the disease. Then, childs, age six, eight, nine, 10, 12, they develop pathological albuminuria. This, it's very important to remember that we need to assess the urinary albumin to creatinine ratio to assess pathological albuminuria, because another early feature Fabry nephropathy is polyuria-- inability to concentrate urine. So if we just assess urinary protein by dipstick, this may become negative because they have high amount of urine, because anything in urine will be diluted, including proteins.

So in childhood, you already have pathological albuminuria, what used to be called microalbuminuria. This slowly increases over the years, over the teenage years, over their 20s, until it reaches overt proteinuria level. That's albuminuria over 300 of milligrams per gram of urinary creatinine. Proteinuria will further increase until it reaches-- it may reach even nephrotic range.

So proteinuria may increase up to the nephrotic range, over 3 grams of ordinary protein per day or per gram of urinary creatinine. But it will never give rise to nephrotic syndrome. It will never give rise to nephrotic syndrome. Patients with nephrotic syndrome, even if they have a prior diagnosis of kidney disease, they should be suspected to have some additional kidney disease.

Eventually, at age 30, 35, glomerular filtration rate will start to decrease at quite a fast rate-- around 5 to 10 mls per minutes per year-- very similar to the rate of loss of glomerular filtration rate in diabetic nephropathy. Patients will need dialysis at a mean age of around 40 years old.

So overall, we have this very long natural history, around 40 years. When they are born, they already have deposits in kidneys cells. In childhood, they develop pathological albuminuria. In early adulthood, they develop overt proteinuria, up to nephrotic range proteinuria. At around age 40, they will need dialysis. 40 years natural course.