Abstract: Background - Denys Drash syndrome is a very rare disorder characterized by gonadal dysgenesis, nephropathy and Wilms tumor. Case report - A 1 year 6 months boy presented with bilateral undescended testes, mid penile hypospadias and abdominal mass. On evaluation found to have Wilms tumor, for which left nephrectomy was done. He was treated with chemo radiotherapy post operatively. Orchidopexy of both testes was done in stages following which he developed nephrotic syndrome resistant to steroids. Thus Denys Drash syndrome was made out. He is on Mendoza regimen currently for SRNS. He has completed chemotherapy and is on regular follow up for Wilms tumor and SRNS and is now 3 years. 

Keyword: Undescended testes, Wilms tumor, Denys Drash syndrome, Steroid resistant nephritic syndrome

The Child with Scar for Nephrectomy and B/L Orchidopexy. Also note short penis with small testis.

Background
Denys Drash syndrome is characterized by diffuse mesangial sclerosis, male pseudohermaphroditism, 46XY karyotype and nephroblastoma. It is due to mutations in WT1 gene.

CASE REPORT
A boy of 1 year 6 months first born to non-consanguineous parents presented with history of abdominal distension with pain for 3 months duration. There was history of decreased appetite. There was no history of fever, altered sensorium, visual disturbances or edema. There is no similar history in the past. Family history was not suggestive of any renal disease or death in early age. On examination he had pallor, no edema. He had fullness in left lumbar region and bilateral undescended testes with mid penile hypospadias. He was normotensive. Abdominal examination revealed left kidney enlargement. Other system examination was clinically normal. Investigations were done. RBC casts were seen in urine microscopy. Ultrasound abdomen was suggestive of left nephromegaly with altered echoes and a normal right kidney. Both testes were in inguinal region. Renal parameters were elevated. Karyotyping was done and it was XY. Left nephrectomy was done and the histopathological examination suggestive of Wilms TUMOR with triphasic pattern with blastemal, stromal and epithelial pattern and a favorable histopathology and no evidence of metastasis to lymph nodes. He was staged as Stage 2 but upstaged to 3 and was treated with post operative chemotherapy with vincristine, dactinomycin, and doxorubicin followed by radiotherapy. On follow up ultrasound abdomen there was no residual mass on left side and right kidney of normal size with grade 1 echoes. After three months of left nephrectomy, left orchidopexy was done. The child had right inguinal hernia for which right herniotomy and right orchidopexy was done after 7 months of nephrectomy. Post operatively the child developed peri orbital puffiness, pedal edema, hypertension. Urine examination revealed nephrotic range of proteinuria. He was initially treated with prednisolone for which he did not respond for 8 weeks of full dose steroid therapy. Now he is on Mendoza regimen for which he did not respond for 8 weeks of full dose steroid therapy. Now he is on Mendoza regimen. The child is thus diagnosed to have DENYS-DRASH SYNDROME presented with Wilms tumor and steroid resistant nephrotic syndrome (SRNS). The child is now receiving IV pulse doses of methyl prednisolone for SRNS as a part of Mendoza regimen but has not attained any remission. He is also on regular follow up USG for imaging kidneys (for Rt kidney Wilms tumor risk) and gonads (risk for gonadoblastoma) and regular renal function tests with urine for proteinuria. His parents did not consent for a renal biopsy and hence it was not done. They also did not decide yet upon gonadectomy after counseling. His renal function tests are now normal except for proteinuria. He is now on Enalapril to control proteinuria and hypertension. Mutation analysis for exon 9 of WT1 gene alone was done and it was negative. Other exons were not analysed.

Discussion:
Denys-Drash syndrome is a condition that affects the kidneys and genitalia. Denys-Drash syndrome is characterized by kidney disease that begins within the first few months of life. Affected individuals have a condition called diffuse glomerulosclerosis, in which scar tissue forms throughout glomeruli, which are the tiny blood vessels in the kidneys that filter waste from blood. In people with Denys-Drash syndrome, this condition often leads to kidney failure in childhood. People with Denys-Drash syndrome have an estimated 90 percent chance of developing a rare form of kidney cancer known as Wilms tumor. Affected individuals may develop multiple tumors in one or both kidneys. Although
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