Movat's (1961) method, these humps can be visualized using both optical and electron microscopical methods. They can also be demonstrated on renal biopsy material prepared for light microscopy using minor modifications of a chromotrop-silver methenamine method (T. Ehrenreich and T. Espinosa, 1967, personal communication). These appearances were observed in renal biopsy specimens from 6 out of 7 children diagnosed as having acute post-streptococcal glomerulonephritis, but with atypical clinical features, and in one child who had had the initial streptococcal infection 17 months before biopsy. Humps were also identified in renal biopsy and necropsy material from a child with rapidly progressive glomerulonephritis, a condition in which they have not previously been described.

**Reference**


Dilatation of the urinary tract may be due either to anatomical obstruction or to dysplasia; it is sometimes difficult to distinguish between these by radiological techniques and to decide whether significant obstruction requiring surgery is present. The ideal pre-operative assessment would predict the improvement in renal function to be obtained from surgical intervention.

Studies of experimental unilateral hydropnephrosis in dogs (Suki et al., 1966) indicate that a smaller fraction of filtered water (V/GFR: V = urine flow rate, GFR = glomerular filtration rate) is excreted during water diuresis by an acutely obstructed kidney than by its control kidney. In contrast, V/GFR is increased in chronic obstruction. It might be expected that those kidneys with the functional characteristics of the acutely obstructed model would show the greatest increase in GFR after relief of obstruction.

GFR was estimated from the plasma disappearance of 51Cr-EDTA (Chantler et al., 1969) before and after surgery in children with presumed obstructive uropathy. Fractional water excretion during pre-operative water diuresis was measured as the plasma/urine creatinine ratio (Pc/Uc):

\[
\frac{V}{GFR} = \frac{U_cV}{P_c} = \frac{P_c}{U_c}
\]

Fourteen children have been satisfactorily examined to date: the largest fractional increases in GFR after surgery have been seen in the three children who had values of P_c/U_c during water diuresis lower than healthy adults. The other 11 had values higher than those of healthy adults.

**Reference**


**Adrenocortical Atrophy and Diffuse Cerebral Sclerosis: Biochemical Studies.** Constance C. Forsyth (Department of Child Health, University of Dundee). A boy, diagnosed as having Addison's disease due to idiopathic atrophy of the adrenal glands at the age of 7 years, developed the first evidence of what was originally thought to be Schilder's disease at 8 years and 10 months. He died at 9 years and 11 months. There are 12 fully documented reports in the literature of boys with the combination of adrenocortical atrophy and diffuse cerebral sclerosis. The studies of adrenal function during life and of the biochemistry of the brain at necropsy were discussed in relation to theories of causation and the likely mode of inheritance of the condition.

**Enzyme Studies of the Testis in Myotonic Dystrophy.** William Hamilton and R. G. Mitchell (Department of Child Health, University of Aberdeen).

Myotonic muscular dystrophy is characterized by familial incidence, inheritance as a dominant trait with variable penetrance, myopathy mainly involving the distal musculature of the extremities and the cranial muscles, cataracts, bony lesions, and endocrine defects such as a low BMR, progressive testicular atrophy, and low urinary 17-oxosteroids.

The testes show tubular degeneration with sclerosis and hyalinization, but Leydig cells may be increased. Testicular biopsies from two affected brothers have been studied. The specimens were incubated as described by Hamilton, Grant, and Wilson (1970). These authors observed that fetal testes synthesized testosterone preferentially from dehydroepiandrosterone before the 12th week of gestation but after that time, preferentially from androstenedione. Undescended testes behaved in vitro like the later fetal testes.

The corresponding enzymic activity of testes from these two boys is set out in the Table:

<table>
<thead>
<tr>
<th>TABLE</th>
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<tbody>
<tr>
<td>Conversion to Testosterone (dpn) as % of Substrate Used</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Elder 13 years</td>
</tr>
<tr>
<td>Younger 11 years</td>
</tr>
</tbody>
</table>

Since the conversion of androstenedione to testosterone is by 17β-reductase, the findings might indicate that as the disease progresses there is progressive failure of 17β-reductase. Also dehydroepiandrosterone was utilized more by the older testis than by the younger testis. The enzyme in this reaction is 3β-hydroxysteroid dehydrogenase which is also the more active enzyme in the early fetal testis. Its persistence then as the
Adrenocortical atrophy and diffuse cerebral sclerosis: biochemical studies.

C C Forsyth

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