Henry Ford Hospital Medical Journal

Volume 1 | Number 3 Article 6

9-1953

Recent Therapy of Nephrosis: A report of fifty-eight cases

Bernard A. O'Hora

RECENT THERAPY OF NEPHROSIS

A report of fifty-eight cases.

BERNARD A. O'HORA, M.D.*

The nephrotic syndrone in children has gained new interest with the introduction of antibiotics for the control of intercurrent infection, and, more recently through the use of ACTH in inducing diuresis and remission of symptoms of this disease. The nephrotic syndrone has long been characterized, and in the minds of most, differentiated from glomerulonephritis—by insidious onset, quite unlike that of glomerulonephritis, by the lack of impairment of renal function, by ability to resolve with complete recovery and by the unique susceptibility of its victims to pneumococcal bacteremia and peritonitis. It is this last feature which was responsible for the fatal outcome of most cases prior to 1937. In an effort to assess the present prognosis in lipoid nephrosis, our experience is here reviewed.

The well known disease of nephrosis is characterised by edema, albuminuria, low total blood proteins, especially the albumin, normal non-protein nitrogen, normal blood pressure, and absence of gross hematuria. The elevated blood lipids, especially cholesterol and lipoid bodies in the urine have lead to the associated term lipoid nephrosis.

The material studied consists of fifty-eight consecutive children diagnosed as lipoid nephrosis, or the nephrotic syndrone, admitted to the in-patient department over a period of twenty-five years from 1928 through 1953. All of these were, at the time of first admission, clinically and chemically "pure nephrotics" without evidence by history or laboratory determination of glomerulonephritis.

The group consists of twenty-three females and thirty-five males, ranging in age, at the onset of symptoms, from eleven months to nine years. The mean age of onset differed but slightly for the two sexes being thirty-seven months in the females and thirty-eight months in the males. More than eighty-five per cent of the group were between one and five years of age at the onset of symptoms.

It has been possible to follow forty-one patients, all but seventeen of the children, through the course of the disease to eventual recovery or death. Of the seventeen, ten are active cases still having symptoms of the disease and seven have failed to report for follow-up examinations.

The over-all results to date (8-1-53), may be summarized as follows: in Table 1 and Table 2.

Table 1—PRESENT STATUS OF FIFTY-EIGHT NEPHROTIC CHILDREN

	Number	Percent
Recovery	18	37.9
Mild Albuminuria only	4	
Died—Infection	7	32.7
Died—Uremia	12	
Lost to Study	7	12.1
Active, under Therapy	10	17.3
Totals	58	100.

*Resident—Department of Pediatrics.

Table 2—PRESENT STATUS OF THE FORTY-ONE CLOSED CASES

	Number	Percent
Recovered	18	53.7
Mild Albuminuria	4	
Died—Infection	7	46.3
Uremia	12	

From this it will be seen that slightly more than half of the cases, followed to conclusion, may be classified as recoveries. The advent of chemotherapy and antibiotics has altered the prognosis somewhat since all three of our deaths prior to 1937 were due to pneumococcal peritonitis. Since that time only four have died of infection, two of peritonitis, one of lobar pneumonia following varicella, and one of recurrent pleurisy with effusions. The more recent deaths have been largely due to renal failure and have occurred primarily in those cases showing a "mixed" type of picture with a superimposed element of glomerulonephritis. As will be described, such involment has carried a very poor prognosis in our series.

Hematuria is not part of the usual picture seen in the "pure nephrotic" and when it occurs is usually interpreted as evidence of so called "mixed nephrosis" or "nephritic nephrosis" with the implication of a bad prognosis. Microscopic hematuria was a frequent finding in this group, appearing in twenty-five cases one or more times during the course of the illness. As an indication of prognostic significance it may be questioned, however, since six of these cases are known to have completely recovered, five are still active, with four of these apparently improving, and only eleven are known to have succumbed to the disease. By temporarily eliminating from consideration the remaining three cases in which no follow-up is available, it would seem that microscopic hematuria does not, in itself, carry a bad prognosis in the nephrotic child and must be assessed as a quantitative sign.

It is worthy of note that the same statement cannot be made when hematuria accompanies hypertension or azotemia, and this was the case in all eleven who succumbed, while those who survived demonstrated hematuria as an isolated complication.

Hypertension, as a complication is strongly indicative of a superimposed glomerulonephritis and carries a poor prognosis. Seven children in this series have shown definitely elevated blood pressures, a systolic of over 134 mm. Hg. and a diastolic of over 89 mm. Hg. Of this group only one has definitely recovered, one survives as an active case in poor condition and five are dead, uremia being the cause of death in all.

Azotemia is of even more serious prognostic importance. Fifteen children in this series have exhibited blood non-protein nitrogens of 50 mgm. per cent or over, and of these thirteen are dead, one is active and in poor condition, the same case mentioned above as having hypertension, and the other has been lost to study.

Duration of symptoms from onset to eventual recovery or death has ranged from three months, (two cases, one recovery and one death) to over eight years

(one recovery). The usual course has been eighteen to thirty months in those recovering and twenty-four to thirty-six months in those terminating fatally.

Therapy has varied widely over the twenty-five year period covered in this series and it is difficult to credit any single measure, excepting the antibiotics, with contributing to an improved prognosis. General measures used in the group have included transfusions, plasma, concentrated plasma, salt-free albumin, cevitamic acid, thyroid extract, forced high protein diets to the point of leaving in place a polyethylene tube, and low salt diets. The antibiotics have assuredly reduced the mortality due to intercurrent infection, so prevalent in this disease. It might also seem that they would also be of benefit in preventing superimposition of glomerulonephritis, but in our experience this has not been the case. At least four of our uremic deaths have occurred in patients who developed "mixed nephrosis" while on prophylactic antibiotics or chemotheraputic agents. The mechanism here is obscure but it would appear that it is not identical to the pattern usually seen in the development of acute glomerulonephritis following streptococcal infection.

The introduction of adrenal corticotrophic hormone (ACTH) has opened a new phase in the management of nephrotic children. In our series fifteen children have been given a total of twenty-six courses of ACTH. The over-all results of these have been diuresis with remission of symptoms in seventeen instances involving ten children. In one instance the remission was permanent with no recurrence of symptoms and negative urine findings twenty-six months after treatment. A second case has had no edema for thirty-nine months following a course of ACTH but still has a mild albuminuria. Remissions in the other cases have been only temporary. One child has had diuresis following each of four courses of ACTH over a fifteen-month period. Most of our cases not responding to ACTH were among the early cases treated with this drug and were given inadequate dosages in the light of more recent experiences. Our current program is based on the use of forty units of ACTH per day in four doses for a period of ten days. The presence of hypertension or azotemia contraindicates the use of ACTH in our hands. There have been no untoward effects noted to this program.

SUMMARY

1. Fifty-eight cases of Nephrosis seen in the Department of Pediatrics over the past twenty-five years are reviewed.

2. Slightly better than one-half of those patients, twenty-two of forty-one followed for the entire course of the disease, experienced clinical recovery.

3. The development of transient microscopic hematuria by nephrotic children, in the absence of other complications, does not alter prognosis.

4. Development of azotema and/or hypertension is of serious prognostic import.

5. Antibiotic and chemotheraputic agents, have greatly reduced mortality due to intercurrent infections to which nephrotic children are peculiarly prone.

6. ACTH has proven of substantial value in inducing temporary, and in some few cases apparently permanent, remission of symptoms in the nephrotic syndrone.