A case of non-parasitic chyluria in a Bahraini female patient is presented. The aetiology, clinical presentations, pathological findings and management of chyluria are reviewed.

CHYLURIA is the passage of milky or creamy urine due to the presence of chyle. Albumin, fat and fibrin in varying proportions are the chief constituents of chyle. The condition was described several centuries ago by Hippocrates, Galen and Theophile and only recently the mechanism of its dynamics were clearly understood. It is classified into two main types; (a) Parasitic group recognized very commonly in the tropics and sub-tropics as a result of obstruction of the lymphatics draining the urogenital tract due to Wuchereria bancrofti (16). Other parasites such as Echinococcus, Cysticercus cellulosea, Ascaris, Tinea and malaria parasites have also been incriminated (16). (b) Non-parasitic group in which no parasitic factor is involved (8) and clinically present itself as a metabolic disturbance due to loss of lipids (10) or more rarely associated with obstructive symptoms. This paper describes the first case of non-parasitic chyluria diagnosed in Bahrain and to the best of our knowledge the first case in the Arabian Gulf region as well. The possible aetiological factors, clinical presentation, diagnostic criteria and the methods and results of treatments will also be discussed.

Non-Parasitic Chyluria *

By E. Amin ** F. Al-Hilli ***

CASE REPORT

In November 1975 a 20 years old Bahrain housewife presented at Salmaniya Medical Centre complaining of right side loin pain and the passage of milky white urine and occasional haematuria. There was no significant past history of note nor any family history of similar complaint. The patient was healthy and examination and laboratory investigations at that time showed no abnormality except for the presence of chyle in her urine. The symptoms increased remarkably during her third pregnancy in April 1976 especially on rising in the morning. Her symptoms recurred in July 1978 when it was noticed that the urine was sterile but contained traces of protein and few red blood cells. Her IVP was normal. In July 1979 she was diagnosed in India as having proliferative glomerulonephritis and was given immunosuppressive and steroid therapy but with no improvement. In January 1980 she was seen again in Salmaniya Medical Centre with the same complaint but the haematuria was more frequent. The patient was healthy looking with no abnormality on physical examination except some tenderness on right loin. The possibility that the milky urine is associated with filariasis was considered but microbiological investigation of nocturnal specimens failed to demonstrate any abnormality. Her urine was sterile and negative for tubercle bacilli but was heavily loaded with protein and contained many red and white cells as well as epithelial cells. All other laboratory investigations were within normal limits. Cystoscopy showed that both ureteric orifices were secreting milky urine more on the right side. This was followed by transpedal lymphangiography (Figure 1) which confirmed the presence of lymphatico-calyceal fistulations on both sides more marked on the right side. A number of widely dilated lymph vessels in the renal hilum were also seen on both sides. The thoracic duct (Figure 2) was patent and well outlined with the dye indicating that no obstruction existed to the normal flow of chyle. On January 13, 1980 the right kidney was explored and the lower polar artery was preserved. The renal pedicle was dissected and the large lymphatic vessels were exposed particularly around the renal vein.

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** Department of Surgery, Salmaniya Medical Centre, Bahrain
*** Department of Pathology, Salmaniya Medical Centre, Bahrain

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anteriorly. These vessels were mobilized, ligated and divided. The post-operative period was uneventful and the patient was discharged on the 10th day. She was followed on the out-patient clinic. Her loin pain has disappeared and the urine was chyle free most of the time. On June 22, 1980 an IVP was done and she underwent stripping of left renal pedicle to disconnect the lymphatico-calyceal fistulation on the left side. The patient is now free of symptoms.

Histological examination of the disconnected pedicle lymphatics of both sides revealed several widely dilated thick-walled lymphatic vessels with no evidence of filariasis. (Figure 3).

DISCUSSION
Two theories were put forward to explain the aetiology of chyluria. In 1840 Prout (12) originated the secretory theory and suggested that fat was secreted from blood stream into the kidney. This theory which substantiated that increased fat intake leads to chyluria failed to explain the effect of posture and

(Figure 1) Lymphangiogram showing widely dilated lymph vessels in the renal bilum and lymphatico-calyceal fistulations on both sides.

(Figure 2) Lymphangiogram showing patent and well outlined thoracic duct indicating that no obstruction existed to the normal flow of chyle.

(Figure 3) Microsection of the disconnected dilated lymphatic pedicle of renal hilum.
exercise on chyluria. Carter in 1862 (1) evolved the obstructive theory explaining chyluria on the basis of mechanical obstruction between the intestinal lacteals and the termination of the thoracic duct. As a result the obstruction would produce an increased intralymphatic pressure, lymphangiectasis, vascular incompetence and retrograde flow of chyle and lymphatico-renal shunt. Rupture of the lymphatics into the kidney and the establishment of the lymphaticourinary fistulation has been explained on the basis of a relative lack of supportive tissue elements in the renal parenchyma associated with inadequate supply of the collateral blood vessels.

The lymphatic vessels in the embryo develop from primitive lymph sacs of which two lie in the perirenal area. The dilated lymphatics described in the present case and demonstrated by lymphangiography may represent an abnormal or incomplete differentiation of these sacs. In support of such congenital hypothesis is the absence of obstruction to these lymphatics and to the thoracic duct.

The disease is usually unilateral with the left kidney more frequently affected (15). It manifest itself in the second or third decade (11) and often associated with renal tuberculosis, ureteric stones, renal cysts, carcinoma of renal pelvis and hydrocele (17, 18). A variety of other possible aetiological factors have also been suggested which could produce obstruction of the thoracic duct or contributing lymphatics. These include malignant neoplasm of the retroperitoneum, abscesses, diabetes mellitus, mechanical processes (pregnancy) and trauma (8).

The demonstration of fat in the urine in the form of minute molecules or fine needle-like structures confirm the diagnosis of chyluria. But the absence of fat in a single random urine sample in a patient who has had a history of milky urine does not rule out chyluria nor it does mean that the disease is cured. It is well known that remission is common and may be for long time (18). In the present case the patient has had repeated remission for 5 years. Once the diagnosis is made, the approach must be directed towards assessing: (a) Possible aetiological factors. (b) Severity of the condition. (c) Site of lymphatico-calycal fistulation. The severity of the disease could be guided by the signs and symptoms particularly the persistence of symptoms, presence of chylous clots in the urine and loss of body weight due to loss of lipids (18). The formation of fibrinous clot with its irritative effect in the genitourinary tract accounts for symptoms of backache, urgency, frequency, renal colic and rarely acute urinary retention (5, 6, 15, 17). Haematuria may lead to iron-deficiency anaemia (16) and massive proteinuria may cause hypoalbuminaemia and reversal of Albumin/Globulin ratio. Recurrence and exacerbation have been attributed to such factors as posture, exercise, diet, menstruation and pregnancy (16).

Attention must be focussed on all possible aetiological factors that may produce derangement in the retroperitoneal lymphatics. These can be investigated along the following lines: (a) Detection of microfilaria in nocturnal blood smears. (b) Absolute eosinophilic count to detect possible parasitic infestation. (c) Urine for acid fast bacilli. (d) Histological examination of retroperitoneal lymph nodes and lymphatics (18) which may reveal filarial segments or evidence of inflammation and fibrosis.

Non-parasitic chyluria must also be differentiated from severe pyuria and lipuria. The microscopic examination of urine in pyuria will reveal pus cells while in lipuria large fat globules rather than minute molecules or fine needle-like structures are seen. Moreover, lipuria, unlike chyluria show no fibrin and the patient clinically suffer from diabetes mellitus or phosphorus poisoning (8).

Transpedal lymphangiography is essential to demonstrate the site of lymphatico-calycal fistulation (3, 17, 18). Cystoscopy however, during remission of acute symptoms is helpful in determining the affected side of the urinary tract (17, 18).

In general no treatment of chyluria is necessary if there is no general disturbance in health (9). Spontaneous remission has been reported (11). Conservative management must always be encouraged in mild forms of chyluria. This include fat free diet (13) and its substitution with medium chain triglycerides (7). Piperadine derivatives has been used but these are effective in the treatment of Wuchereria bancrofti and their effect is not clear for chyluria (11). But in severe forms, surgical intervention is indicated to prevent progressive debilitation from excessive loss of absorbed nutrients as well as to avoid damaging the upper urinary tract from chylous clot obstructive uropathy (18). Various surgical procedures have been described including nephrectomy, renal capsulectomy, retrograde injection of sclerosing material, local diathermy of the fistula and anastomosis of a varicose lymphatic and the testicular or ovarian vein (4, 14, 16). However, the meticulous dissection and disconnection of the renal pedicle lymphatics i.e. disconnection of lymphatico-calycal fistulation is
the only satisfactory method of treatment (2, 10, 15, 17, 18). In this stripping procedure, the retroperitoneal approach is preferred to transperitoneal route as better mobilisation of the kidney is possible and more meticulous skeletonization of the renal pedicle is achieved (18). Should however, both kidneys are involved as the present case it is advisable to operate first on the side of severe leakage and followed after few months by stripping of the other side. This is because; (a) The symptoms may subside after unilateral intervention. (b) Permanent derangement of kidney dysfunction will not be expected. (c) Simultaneous bilateral stripping invariably leads to incurrence of symptoms.

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