intravenously daily. Just when improvement was being anticipated, the patient collapsed with a myocardial infarct. Bronchopneumonia supervened and in spite of every care he died. Post mortem examination revealed no signs of pemphigus other than in the conjunctiva.

SUMMARY

Ocular pemphigus is reviewed and the possibility that it is a separate entity from pemphigus of other regions is discussed. One case of pemphigus of the conjunctiva only is presented.

622 Medical-Dental Bldg.

REFERENCES

1. CHURCH, R. E. AND SNEDDON, I. B.: *Brit. J. Dermat.*, 65: 235, 1953.
2. LEVER, W. F.: *A. M. A. Arch. Dermat. & Syph.*, 64: 727, 1951.
3. CIVATTE, A.: *Ann. dermat. et syph.*, 3: 1, 1943.
4. LEVER, W. F.: *Am. J. Orthodontics*, 28: 569, 1942.
5. FRANKE, E.: Der Pemphigus und die essentielle Schrumpfung der Bindehaut des Auges, J. F. Bergmann, Wiesbaden, 1900.
6. POSEY, W. C.: *Am. J. Ophth.*, 3: 507, 1920.
7. KING, M. J.: *Am. J. Ophth.*, 16: 903, 1933.
8. WEILL, E. AND DUFOURT, A.: *Lyon. Méd.*, 118: 990, 1912.
9. HARDY, W. F. AND LAMB, H. D.: *Am. J. Ophth.*, O.S. 34: 289, 1917.
10. LEVER, W. F. AND TALBOTT, J. H.: *Arch. Dermat. & Syph.*, 46: 348, 1942.
11. KLAUDER, J. V. AND COWAN, A.: *Am. J. Ophth.*, 25: 643, 1942.
12. SOUDAKOFF, P. S. AND WHALMAN, H. F.: *Am. J. Ophth.*, 36: 231, 1953.
13. MACHT, D. I. AND OSTRO, M.: *Urol. & Cutan. Rev.*, 51: 651, 1947.
14. RYCROFT, B. W.: *Brit. J. Ophth.*, 18: 571, 1934.
15. KLAUDER, J. V.: *Ibid.*, 37: 650, 1938.
16. KANEE, B.: *Arch. Dermat. & Syph.*, 55: 37, 1947.
17. CORBOY, P. M.: *Am. J. Ophth.*, 34: 1561, 1951.
18. KAPUSCINSKI, W. J.: *Ann. ocul.*, 174: 451, 1937.
19. FRIEDMAN, M. W. AND WRIGHT, E. S.: *Am. J. Ophth.*, 36: 237, 1953.

IDIOPATHIC GALACTOSE INTOLERANCE IN A PREMATURE INFANT*

JOHN C. RATHBUN, M.D., F.R.C.P.[C.],†
London, Ont.

SINCE VON REUSS[1] in 1908 reported the case of a child who was unable to metabolize galactose, the literature has gradually increased until today there are 30 proven reports of idiopathic galactose intolerance. Many others, incompletely reported, are to be found in ophthalmological journals.

The condition is characterized by failure to gain in weight, frequent episodes of vomiting and diarrhœa, enlargement of the liver, galactose intolerance as evidenced by abnormal blood galactose tolerance curves and galactosuria, and frequently by mental retardation and cataracts.

The literature up to 1950 was carefully reviewed by Townsend *et al.*[2] and during the past four years 16 more cases have been completely reported.[3-14] Many others are referred to at various meetings and the condition is much more common than the reports would imply. A careful review revealed only one case in a premature infant, reported by Gorter[15] to the British Pædiatric Association in 1951, and this baby died during the fourth day of life. The present case is presented from interest in the possible relationship between prematurity and galactose intolerance.

CASE REPORT

J.C., a female infant weighing 1,956 gm., was born on April 13, 1953, by spontaneous delivery at 8 months' gestation. She was a fairly well developed premature infant who appeared quite normal for her size. She was placed on a diet of 2/3 breast milk and 1/3 protein milk, which she took fairly well. During the next two weeks she failed to gain weight and was admitted to War Memorial Children's Hospital on April 25. At this time her abdomen was full in appearance and the liver was palpable and firm, 1.5 cm. below the costal margin. The tip of the spleen was just palpable. The navel was mildly infected with coagulase-positive *Staphylococcus pyogenes aureus*. Rigorous treatment with antibiotics was given but the skin infection spread, with pustules developing in the scalp. The child was very irritable. By May 4 the liver margin had descended to 2.0 cm. below the costal margin. Her hæmoglobin value had fallen to 9.7 gm. %, and a transfusion of whole blood was given. On the following day, the anterior fontanelle was full and there was some head retraction. The spinal fluid was, however, within normal limits for cells and protein. She began to run a septic temperature, up to 102° F., with a white cell count ranging from 6,600 to 12,700 per c.mm. and a differential of about 50% polymorphonuclear leucocytes. Her weight remained constant, although the child was growing in length, with the result that skin folds were hanging about her upper thighs (Fig. 1). By May 13, it became apparent that infection alone was not the cause of all the difficulty. Investigation of her urine showed a persisent 3-plus qualitative Benedict test, and the sugar was rapidly identified as galactose by osazone crystallization and paper chromatography. On May 18, she was placed on a soy bean flour formula and gained immediately and steadily up to 2,580 gm. by May 30, i.e. 766 gm. in 13 days. With this, there was a remarkable improvement in her infection and drugs were discontinued, except for vitamins and iron, on May 25.

Her subsequent course was uneventful except for two transfusions to raise her hæmoglobin value to normal levels. She was sent home on June 13, weighing 2,637 gm. Physical examination at this time revealed normal fundi and media. The liver was palpable 1.0 cm. below the costal margin, and the tip of the spleen could just be felt. The remainder of the examination was essentially negative.

Follow-up.—Her subsequent development has been satisfactory. Foods were added at the usual time, care being taken that no galactose was given. At 6 months she weighed 6,606 gm., and at 15 months she weighed 9,242 gm. By 15 months of age, her liver had returned

*Presented in part to the 15th Congrès des pédiatres de Langue Française, Marseille, May 24, 1955.
†From the wards of the War Memorial Children's Hospital, London, Ontario, and Departments of Pædiatrics and Medical Research, University of Western Ontario.

Canad. M. A. J.
June 15, 1955, vol. 72

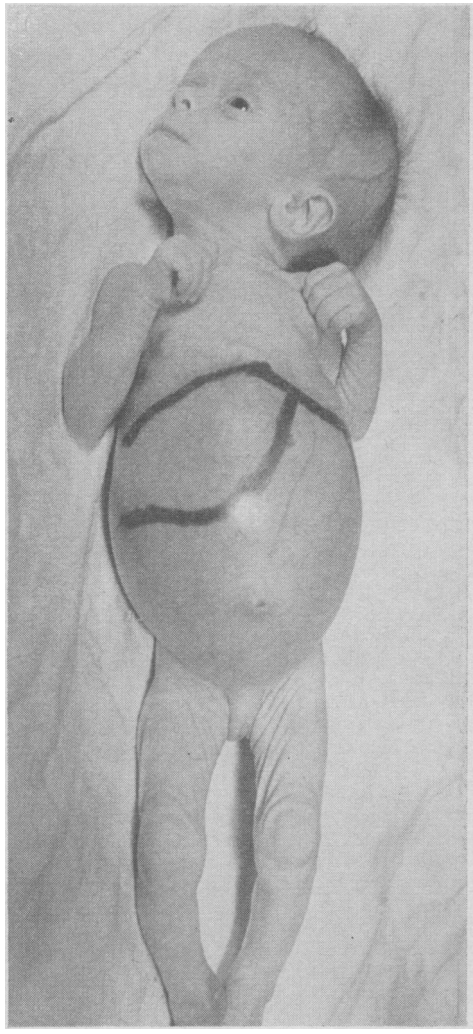

Fig. 1.—Age one month.

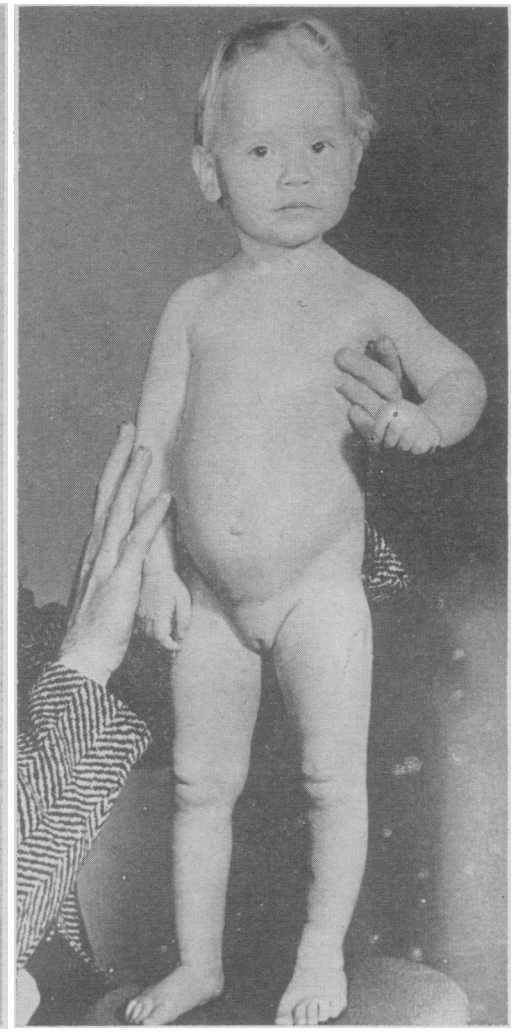

Fig. 2.—Age 15 months.

TABLE I.

BLOOD LEVELS FOLLOWING THE INTRAVENOUS INJECTION OF 1.2 GM. OF GALACTOSE PER KG. OF BODY WEIGHT

Subject	*Age (mos.)*	*Body weight (Kg.)*	*Blood levels galactose (mgm./100 c.c.) Time*			
			0′ blood blank	*15′*	*45′*	*75′*
J.C.—Patient	15	9.3	19	259	213	179
D.E.—Convalescent control.....	11	9.0	17	235	83	31

to normal size, 0.5 cm. below the costal margin, and her spleen was no longer palpable. Her appearance was that of a normal child (Fig. 2).

On July 16, 1954, an intravenous galactose tolerance test was performed, in accordance with the method previously outlined.[16] Galactose was determined in the blood by the method of Fister.[17] As a control, a similar test was performed on a healthy convalescent infant. The results are shown in Table I.

DISCUSSION

Galactose intolerance has been suggested to be either an inborn hereditary metabolic defect by Donnell and Lann and others,[2, 3, 15] or a congenital disturbance in bile excretion by Edmonds, Hennigar and Crooks.[6] In either case, the cause would appear to be active *in utero*. The signs of intolerance appear early and Gorter[15] has reported cataracts present at four days of age. Townsend *et al.*[2] found galactose in the urine of one of their patients as soon as an evaporated milk formula was started, where a family history of galactose intolerance caused a careful watch for galactose to be kept.

Lockhart and Roboz[14] suggested that galactose, transferred by the placental circulation from the maternal blood, is excreted by the fetal kidney and found in the amniotic fluid. These factors suggest that this syndrome is existent

in utero, and one would therefore expect signs of failure to gain with a low birth weight. A review of the literature and all available birth weights gives an incidence curve as shown in Fig. 3. The unimodal peak corresponds to 3,001-3,500 gm., which is identical with that reported

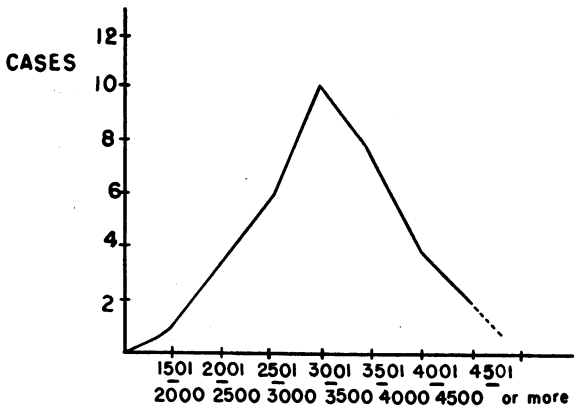

Fig. 3.—Birth weight in grams. Variation in birth weights of reported cases.

by Wegman,[18] quoting the National Office of Vital Statistics of the U.S.A., for all live births in the U.S.A. during the first quarter of 1950. This suggests that the birth of a premature infant with this complaint is an accident and unrelated to its carbohydrate metabolic difficulties. Furthermore, many of these children do not show much feeding difficulty for the first two or three weeks; only then is there a gradually increasing amount of trouble, as in the case reported above. Apparently the small amount of galactose present in the maternal blood causes little damage, and it is only when larger amounts of galactose are given by mouth that clinical signs and symptoms appear.

The intravenous galactose tolerance curves of both patient and control were identical with those previously reported.[16]

Several writers[2, 3, 6] have described infection in these cases and the diagnosis has been confused with sepsis, as above. In view of this frequency of infection, it would seem probable that this parallels the frequency seen in diabetes mellitus, where persistent hyperglycæmia is often accompanied by bacterial invasion. The parallel extends further when one considers the cataracts due to galactosæmia and the mental retardation probably due to hypoglycæmia.

The syndrome has many names, of which the best known is galactosæmia. As this refers to a sign only, the names of idiopathic galactose intolerance or galactose diabetes would appear more appropriate.

SUMMARY

1. The case of a premature infant with idiopathic galactose intolerance is reported.

2. There is no apparent relationship between this disease and prematurity.

3. The effect of circulating maternal galactose upon the intrauterine fetus appears to be minimal.

4. Infection is a common finding while hypergalactosæmia is present.

The writer is grateful to Dr. J. A. F. Stevenson and Dr. H. A. DeLuca for the blood galactose studies.

REFERENCES

1. VON REUSS, A.: *Wien. med. Wchnschr.*, **58**: 799, 1908.
2. TOWNSEND, E. H. JR., MASON, H. H. AND STRONG, P. S.: *Pediatrics*, **7**: 760, 1951.
3. DONNELL, G. N. AND LANN, S. H.: *Pediatrics*, **7**: 503, 1951.
4. DUSHANE, J. W. AND HARTMAN, E. E.: *Pediatrics*, **7**: 679, 1951.
5. LANGEWISCH, W. H. AND BIGLER, J. A.: *Pediatrics*, **9**: 263, 1952.
6. EDMONDS, A. M., HENNIGAR, G. R. AND CROOKS, R.: *Pediatrics*, **10**: 40, 1952.
7. BRAY, P. T., ISAAC, R. J. AND WATKINS, A. G.: *Arch. Dis. Childhood*, **27**: 341, 1952.
8. JOHNS, D.: *A. M. A. Am. J. Dis. Child.*, **85**: 575, 1953.
9. STEINER, M. M.: *Am. J. Ophth.*, **36**: 841, 1953.
10. JOHNSON, J.: *Am. J. Ophth.*, **36**: 1380, 1953.
11. HOLZEL, A.: *Med. Illust.*, **8**: 44, 1954.
12. HUDSON, F. P. *et al.*: *Brit. M. J.*, **1**: 242, 1954.
13. FOX, E. G., FYFE, W. M. AND MOLLISON, A. W.: *Brit. M. J.*, **1**: 245, 1954.
14. LOCKHART, J. D. AND ROBOZ, E.: *Pediatrics*, **13**: 211, 1954.
15. GORTER, E.: *Arch. Dis. Childhood*, **26**: 271, 1951.
16. GREENMAN, L. AND RATHBUN, J. C.: *Pediatrics*, **2**: 666, 1948.
17. FISTER, H. J.: Standardized Procedures for Spectrophotometric Chemistry, Standard Scientific Supply Corp., New York, 1950.
18. WEGMAN, M. E.: *Pediatrics*, **14**: 396, 1954.

DAVID ANDERSON-BERRY PRIZE

A David Anderson-Berry Silver-gilt Medal, together with a sum of money amounting to about £100, will be awarded in 1956 by the Royal Society of Edinburgh to the person who, in the opinion of the Council, has recently produced the best work on the therapeutical effect of x-rays on human diseases.

Applications for this prize are invited. They may be based on both published and unpublished work and should be accompanied by copies of relevant papers. Applications must be in the hands of the General Secretary, Royal Society of Edinburgh, 22/24 George Street, Edinburgh, 2, by March 31, 1956.