Glycogen Storage Disease Type IV, Amylopectinosis

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Classification of the glycogen storage diseases according to the underlying enzyme defect has added considerably to our understanding of this group of diseases. Of the 6 types in which the biochemical abnormality has been characterized, the least common appears to be type IV, amylopectinosis. The first case was described by Andersen in 1952, and the glycogen present in the liver and other organs was shown by Illingworth and Cori (1952) to possess abnormally long outer and inner chains of glucose units. For this reason, Cori (1954) suggested that the enzyme, \( \alpha-1,4\)-glucan: \( \alpha-1,4\) glucan 6-glycosyl transferase, effective in transferring chains of glucose units to form new branches in the glycogen molecule, was absent.

Despite this clear recognition of a new syndrome, few cases have since been reported. A second was diagnosed during life by Sidbury et al. in 1962, and a brief account of a third, also diagnosed in life, was given 3 years later (Mortimer, 1965). Since then, descriptions of 3 more have appeared, 2 of whom were sibs (Holleman, van der Haar, and de Vaan, 1966; Brown and Brown, 1966; Fernandes and Huijing, 1968). In the last 2, absence or gross deficiency of the branching enzyme was conclusively demonstrated by assay.

The present article is concerned with the third case to be reported, of which only preliminary details have previously been given (Mortimer, 1965). Glycogen storage disease was suspected at 8 months of age because of gross hepatomegaly associated with enlargement of the spleen. Investigations of carbohydrate metabolism supported the diagnosis, which was confirmed by an examination of a biopsy specimen of liver. The sparing solubility in water of the glycogen extracted from the liver, and the blue colour of its complex with iodine suggested an amylopectin-like structure, which was proved by a more complete examination.

**Case Report**

A boy, the first child of unrelated parents, was born at term in hospital on March 4, 1964, after a normal pregnancy and delivery, birthweight 3515 g. For the first 5 weeks he was breast-fed, with dried milk supplements, but thereafter he was fully artificially fed. He gained weight satisfactorily till he was 6 months old, when he began to vomit and ceased to gain. At 8 months of age, he was admitted to hospital because of anorexia as well as persistent vomiting occurring at least once every day. He was then noted to be an alert and active, but wasted child, his weight being on the 3rd centile. The liver, which was firm and smooth, was grossly enlarged to the level of the iliac crest. The spleen was also enlarged to 3 cm. below the costal margin. Initial investigations showed a moderately severe acidosis, treated at first with M/6 sodium lactate, to which after 2 days potassium acetate was added. He ceased to vomit, and improved clinically. An oral glucose tolerance test gave normal results, but the subnormal hyperglycaemic response after injection of adrenaline and glucagon pointed to glycogen storage disease, and a biopsy of the liver was taken. He was discharged, having gained a little weight, on a normal diet, but still having an alkali mixture in the form of sodium citrate and potassium acetate.

He was readmitted when he was 13 months old because of increasing enlargement of the liver and spleen. In an attempt to reduce glycogen deposition in the liver, he was given a low carbohydrate, high protein diet. He improved at first and began to gain weight (Fig. 1). The improvement was short lived however, and he developed ascites, with shortness of breath, for which he was again admitted to hospital. He was in addition slightly jaundiced, the liver was still grossly enlarged with increased splenomegaly, the abdominal vessels were prominent, and there was some oedema of the feet. Neither repeated paracenteses nor infusion of salt-free albumin were effective in reducing the ascites or oedema, nor were diuretics any more successful. The child’s condition continued to deteriorate, and he was therefore changed to a low protein diet with increased carbohydrate without effect. A portacaval transposition was performed in the hope of decreasing glycogen deposition in the liver but the child died 24 hours later, in liver failure, when 19 months old. Permission for necropsy was refused.

**Material and Methods**

Glucose-6-phosphatase, glycogen synthetase, debrancher enzyme, phosphorylase, and acid maltase

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were estimated in the liver as described by Hers (1959, 1963), and fructose-1-phosphate and fructose-1:6-diphosphate aldolase activities as recorded previously (Solimano, Burgess, and Levin, 1967). Ornithine transcarbamylase was determined by a modification of the method of Kulhanek and Vojtšíková (1964). The glycogen content, phosphorylase activity, and debrancher enzyme in leucocytes were estimated by methods of Williams and collaborators (Williams and Field, 1961; Williams, Kendig, and Field, 1963). The leucocytes were prepared using a fibrinogen sedimentation technique (Skoog and Beck, 1956).

For the assay of glycogen synthetase, the leucocytes from 15 ml. blood were suspended in 0.1 ml. sucrose—EDTA solution before homogenizing, and the enzyme estimated as in liver.

Blood lactate was estimated by a micro-modification of the enzymatic method of Horn and Bruns (1956), and free fatty acids by a modification of the method of Duncombe (1964).

Glycogen extraction and estimation of glycogen content.

1. Cold trichloroacetic acid method. 0.5 g. liver was homogenized in 5 ml. cold 5% trichloroacetic acid in water. The precipitated protein was centrifuged and extracted with a further 5 ml. 5% trichloroacetic acid.

Glycogen was precipitated from the combined supernatant by the addition of an equal volume of absolute alcohol. After allowing to stand for at least 1 hour at 0-4°C the precipitated glycogen was removed by centrifugation. It was washed first with 75% alcohol and then with absolute alcohol and the residue then dried in a desiccator. The glycogen content of the residue was determined by the anthrone method.

2. Potassium hydroxide method. The glycogen was extracted by adding 1 ml. hot 30% KOH solution to 0.5 g. liver and heating in a boiling water bath for 30 minutes. After cooling, the glycogen was precipitated with 2 ml. absolute alcohol, allowed to stand at 0-4°C for 6 hours, and the precipitate separated by centrifugation. It was washed with absolute alcohol and the glycogen content estimated as before.

Glycogen structure.

Hydrolysis by β-amylase. 0.8 ml. suspension of glycogen containing between 100-500 μg./ml. was incubated at 37°C for 4 hours in 0.1 ml. 2 M acetate buffer pH 5.0, with 0.1 ml. of the suspension of β-amylase (2 mg./ml. in 0.9% saline). After incubation, an aliquot (0.5 ml.) was neutralized with 0.2 N sodium hydroxide solution and 0.2 ml. each of 0.3 N barium hydroxide and 5% zinc sulphate solution were added to precipitate the protein.

The protein-free supernatant (100 μl.) was incubated with 100 μl. maltase suspension (2 mg./ml. in glycylglycine buffer, pH 7.2) for 30 minutes. The solution was centrifuged and the glucose formed was estimated in the supernatant using the glucose oxidase method. A standard curve was obtained using solutions containing 25, 50, and 100 μg./ml. maltose estimated as above.

The absorption spectra of the glycogen-iodine complexes were determined on an aqueous solution of the glycogen preparation, containing 0.2 mg./ml. This was mixed in a cuvette with an equal volume of a solution containing potassium iodide (0.4 g./100 ml.) and iodine (0.04 g./100 ml.). The absorption was read at wavelengths between 400 and 600 mμ at intervals of 20 mμ against a blank containing equal volumes of the iodine/potassium iodide solution and water.

Biochemical investigations.

Apart from a persistently low bicarbonate ion concentration even on treatment, the plasma electrolytes were within normal limits, except during the period before death, when the plasma sodium fell to 120 mEq./l. The blood urea was also normal except after operation when it rose to 103 mg./100 ml. The serum alkaline phosphatase was raised, as were the serum transaminases which remained so throughout his illness. Serum cholesterol was also raised. Initially the total serum protein, albumin, and globulin levels were normal, but as cirrhosis developed, the serum albumin fell and globulin much increased till the few days preceding death, when infusions of plasma and of albumin temporarily restored normal levels. Thymol turbidity, zinc sulphate turbidity, and γ-globulin turbidity were also normal initially, but later they were all greatly increased. The serum bilirubin examined when clinical jaundice became apparent was slightly raised during the later stages of his illness but more severely just before death. The blood ammonia was also increased (69 μg./ml. and 99 μg./ml.) at this stage.
Renal function tests. The hydrogen ion excretion index determined early in the course of the disease was found to be low, 0·8, compared with a normal of greater than 1·2 (Peonides, Levin, and Young, 1966), confirming a renal tubular acidosis. There was neither hyperaminoaciduria nor proteinuria.

Carbohydrate metabolism. There was no evidence of hypoglycaemia throughout the illness, and no ketone bodies were found in the urine except during the glucagon tests. An oral glucose tolerance test on two occasions showed a normal rise in the blood level with normal levels of lactate and free fatty acids (Fig. 2). A fructose tolerance test was also normal, the maximum level of blood fructose being 20 mg. per 100 ml. In neither case did the blood glucose level fall to hypoglycaemic levels at the end of the test (Fig. 3).

Hepatic glycogenolysis. The glucagon test was done on three occasions. The first was when the infant was 8½ months old, 700 µg glucagon being given intramuscularly. The maximum rise in blood sugar was only 14 mg./100 ml., and this occurred as late as 1 hour after the injection. In the second test, when 1 mg. glucagon was given intramuscularly, the maximum rise of blood glucose, 27 mg./100 ml., occurred 30 minutes after injection (Fig. 4a). The third test, 2 hours after a meal high in carbohydrate, showed a similar rise in blood glucose of 25 mg./100 ml. from a high fasting level of 95 mg., again 30 minutes after the injection. The urine passed in the last test contained traces of ketone bodies.

The adrenaline test, also performed twice, gave similar results. The first, in which blood sugar was measured, showed a maximum rise of 15 mg./100 ml. 30 minutes after injection. In the second test (Fig. 4b), the rise in blood glucose was 14 mg./100 ml., also maximal at 30 minutes, with traces of ketone bodies in the urine.

Macrophotical and microscopical appearances of liver. At operation, the liver surface was said to be firm and moderately granular in appearance. The presence of severe portal hypertension was indicated by the grossly dilated porta-systemic anastomoses.

Histology of the liver biopsy showed cirrhosis. The parenchyma cells were enlarged, and the cytoplasm was

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**Fig. 2.**—Oral glucose tolerance test (1·75 g./kg.). Blood lactate and plasma free fatty acid levels are also shown.

**Fig. 3.**—Oral fructose tolerance test (2 g./kg.). Note normal increase of blood glucose level.

**Fig. 4a.**—Glucagon test (1 mg. I.M.). Lower graph shows blood glucose levels when glucagon was injected after 12-hour fast. Upper graph shows results when glucagon was given 2 hours after a high carbohydrate meal.

**Fig. 4b.**—Adrenaline test.
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.. normal rabbit liver glycogen 
- - - Cold TCA extract 
- - - Hot KOH extract 
Patients liver glycogen

Fig. 5.—Absorption spectra of glycogen-iodine complexes. Note peak of patient's liver glycogen (KOH extract) is at 520 mμ compared with peak at 420 mμ for normal glycogen. The curve for the TCA extracted glycogen is between these two.

Isolation of glycogen from liver. The glycogen content of a specimen of liver obtained at operation was determined after extraction with (a) cold 5% trichloroacetic acid, yielding 2.4 g./100 g., (b) hot trichloroacetic acid, yielding 3.6 g./100 g., (c) hot 30% potassium hydroxide solution which gave 4.9 g./100 g. liver.

Structure of liver glycogen. The glycogen obtained by cold trichloroacetic acid extraction differed from that isolated with hot potassium hydroxide in two respects. The former was readily soluble in cold water, and gave a brown-red colour with iodine. The latter was much less soluble and the complex with iodine was blue. Since it was possible that they differed in composition the structures of both were determined.

The absorption spectra of the glycogen-iodine complexes from both specimens are shown in Fig. 5, together with those from (a) normal rabbit glycogen, and (b) glycogen obtained from the liver of a patient with glycogen storage disease type II (Pompe's disease). The maximum absorption of the complex from the glycogen obtained by hot potassium hydroxide was 520 mμ, suggesting that it was similar to that of amylopectin, which gives the same peak, and different from normal glycogen-iodine complex, which has a peak at 420 mμ. The absorption curve by trichloroacetic acid-extracted glycogen showed a plateau rather than a peak (Fig. 5), suggesting the possibility that it was a mixture of amylopectin and normal glycogen.

Hydrolysis of the potassium hydroxide-extracted glycogen by β-amylase resulted in a 66% breakdown to maltose, whereas only 40% hydrolysis occurred from the action of β-amylase on the trichloroacetic acid-extracted glycogen. The mean chain length of the potassium hydroxide-extracted glycogen, measured by the method of Whelan (Lee and Whelan, 1966), was 22, compared with a normal of 11-14. From this result and the degree of hydrolysis by β-amylase, the exterior chain length can be calculated to be 17, compared with a normal of 7-8, while that of the inner branches is 4, compared with the normal of 3-4. The degree of end-group branching is 4-6%, compared with a normal value of 9%. These results suggested a glycogen with abnormally long outer branches, resembling amylopectin (Table I).

<table>
<thead>
<tr>
<th>Case</th>
<th>% Amylalysis</th>
<th>Chain Length</th>
<th>Exterior Chain Length</th>
<th>Interior Chain Length</th>
<th>Branch Points %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present case</td>
<td>66</td>
<td>22</td>
<td>16</td>
<td>4</td>
<td>3.6</td>
</tr>
<tr>
<td>Andersen (1952)</td>
<td>50.5</td>
<td>21.2</td>
<td>14.7</td>
<td>6.5</td>
<td>3.7</td>
</tr>
<tr>
<td>(phosphorylase)</td>
<td>59</td>
<td>--</td>
<td>--</td>
<td>--</td>
<td>--</td>
</tr>
<tr>
<td>Hollman et al. (1966)</td>
<td>57</td>
<td>20.7</td>
<td>14-15</td>
<td>5-6</td>
<td>3.8</td>
</tr>
<tr>
<td>Normal glycogen</td>
<td>38-50</td>
<td>12-14</td>
<td>8-9</td>
<td>3-4</td>
<td>6.7</td>
</tr>
<tr>
<td>Amylopectins</td>
<td>50-60</td>
<td>21-24</td>
<td>14-17</td>
<td>5-8</td>
<td>3.5</td>
</tr>
</tbody>
</table>
TABLE II

<table>
<thead>
<tr>
<th>Liver Enzyme Activities and Glycogen Content</th>
<th>Present Case (μM/g. per min.)</th>
<th>Mean Normal Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glucose-6-phosphatase</td>
<td>5.1</td>
<td>7.4</td>
</tr>
<tr>
<td>Acid maltase</td>
<td>0.89</td>
<td>0-62</td>
</tr>
<tr>
<td>Phosphorylase</td>
<td>13.6</td>
<td>18-7</td>
</tr>
<tr>
<td>Glycogen synthetase</td>
<td>0.94</td>
<td>8-0</td>
</tr>
<tr>
<td>Aldolase F1P</td>
<td>1.4</td>
<td>3-9</td>
</tr>
<tr>
<td>FDP</td>
<td>3.1</td>
<td>6-4</td>
</tr>
<tr>
<td>Ornithine transcarbamylase</td>
<td>37.8</td>
<td>99.1</td>
</tr>
<tr>
<td>Glycogen content (g./100 g.)</td>
<td>4.9</td>
<td>&lt;2</td>
</tr>
</tbody>
</table>

Similar experiments with the trichloroacetic acid-extracted material gave a mean chain length of 16, the exterior branch length being 9, the inner branch length being 6, and the end-group branching being 6·2%, which are abnormal values, though less so than the KOH-extracted material.

Liver enzymes of glycogen metabolism It was unfortunately not possible to estimate the activity of the branching enzyme, since only a small amount of liver was available for study and no purified brancher-free phosphorylase-a was available for the assay. However, glucose-6-phosphatase, acid maltase, phosphorylase, and glycogen synthetase activities in the liver were assayed (Table II). The first three were normal, but glycogen synthetase was much reduced, whether rabbit liver glycogen or the glycogen isolated from the liver of the patient was used as substrate for the assay. Two other enzymes, aldolase and ornithine transcarbamylase, were also normal.

Investigations on leucocytes from parents. The glycogen levels in the leucocytes from both parents were normal (Table III). Though insufficient material was isolated to determine the structure, the glycogeniodine complex was normal in colour and showed a peak only at 420 m_{μ}. Glycogen synthetase, phosphorylase, and debrancher enzyme levels were within normal limits.

Discussion

This disease may not be as rare as is supposed. Andersen (1964) cites a similar case in the older literature, and it seems likely, as Sidbury et al. (1962) suggest, that the 3 cases of polysaccharide storage disease described by Craig and Uzman (1958) were also amylopectinosis.

From the 5 cases including the present one, of which the clinical details are recorded, a uniform clinical picture of amylopectinosis emerges, which confirms the findings in the description of the original case (Andersen, 1952). The affected infant appears normal at birth and for some months thereafter, but early in infancy there is a failure to thrive, and by the sixth month of life abdominal distension due to hepatomegaly, with some splenomegaly, is usually obvious. The child is much below its expected weight, and muscle hypotonia and wasting are also present. As liver and spleen increase in size, the superficial veins over the abdomen become prominent, and ascites develops. Jaundice if present is only slight, being obvious only in the later stages of the illness. Intercurrent infection is frequent, and death occurs from liver failure, in the second year of life, though one child survived to the fourth year. Amylopectinosis may thus be differentiated clinically from the more common glycogenoses, glucose-6-phosphate deficiency, limit dextrinosis, and liver phosphorylase deficiency, which also give rise to hepatomegaly, by the early development of cirrhosis with portal hypertension ending in severe liver failure, and also by the absence of hypoglycaemic episodes.

As in the other forms of glycogenosis, there may be haemorrhagic manifestations in amylopectinosis. These usually take the form of intestinal haemorrhages, presumably due to oesophageal varices. In the present case, purpura was present only in the last stages of the illness when the prothrombin time was greatly increased, and no other haemorrhagic manifestations were noted.

A low carbohydrate diet effected only a short-lived improvement in our case. The effectiveness of portacaval transposition has been claimed in other forms of glycogenosis (Starzl et al., 1965), but it was performed too late in our case to achieve a useful result. In the longest surviving patient (Sidbury et al., 1962), steroids were employed over a long period. More recently, α-glucosidase, a fungal enzyme prepared from Aspergillus niger, has been used by Baudhuin, Hers, and Loeb (1964) to decrease glycogen concentration in the liver in type II glycogenosis. Fernandes and Huizing (1968) have similarly treated amylopectinosis: they recorded a
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4×9 cases, it gen content to an. In the present case, the glycogen content was 4-9 g./100 g. by KOH extraction. Though analysis proved this substance to have a long outer and inner chain structure similar to that of amylopectin, the data for the glycogen obtained by the less drastic extraction process using cold trichloroacetic acid showed that its structure was closer to that of normal glycogen. Thus, the trichloroacetic acid-extracted material was readily soluble in water like normal glycogen, the colour of the iodine complex was similar to that of the normal glycogen-iodine complex, and the percentage hydrolysis by β-amylase was within the range for normal glycogen. The absorption spectrum of the iodine complex of the trichloroacetic acid-extracted glycogen suggests that it is a heterogeneous mixture of glycogen molecules, some with relatively normal branching and some with long branch chains. Evidence in favour of this possibility is also afforded by the appearance, on histological examination, of a centrally placed dense mass of glycogen in the liver cell, with finely granular glycogen particles situated peripherally. Holleman et al. (1966) have also suggested that the stored substances in their case of amylopectinosis were heterogeneous, consisting of a mixture of polysaccharides of different branching characteristics, and evidence based on a fractionation of the glycogen from the liver of their case was adduced in support of this possibility by Brown and Brown (1966).

The absence of the branching enzyme in both liver and leucocytes has now been proved in two affected patients (Brown and Brown, 1966; Fernandes and Huijing, 1968). Though this deficiency explains the formation of an amylopectin-like polysaccharide, it does not account for the presence of an appreciable proportion of branch points in the abnormal glycogen, as Hers (1964) points out. Brown and Brown (1966) suggest that two enzymes, with branching activities of different specificities, are present in normal liver, only one of which is measured by the method used. Another possibility is that the mutant gene in this disorder has produced a protein with substantially modified enzyme specificity, such that branching occurs mainly with long outer chains.

Enzymes of glycogen metabolism. The levels of only 4 enzymes of the glycogen pathway were estimated, and the only abnormality detected was a very low glycogen synthetase activity, only 10% of the mean level found in normal liver. This was not related directly to the abnormal glycogen, since the latter was capable of acting as substrate for mouse liver glycogen synthetase. The glycogen synthetase is sufficiently active in vivo to form glycogen, even if abnormal in structure. However, the very low activity may account for the relatively low liver glycogen content in this con-
condition compared with that found in other glyco-
genoses.

Glycogen synthetase has not been assayed in previous cases of amylpectinosis, and the signifi-
cance of the association of an absent brancher
enzyme with severely diminished glycogen syn-
thetase activity is obscure. However, it may be of
great theoretical interest as multiple enzyme defects
appear to be more frequently reported in glycogen
storage disease than in other inborn errors of
metabolism (Auerbach and DiGeorge, 1961; Stein-
itz, 1967). In these cases the enzymes affected have
usually been a reduced glucose-6-phos-
phatase in association with amylo-1:6-glucosidase
deficiency, but other double enzyme deficiencies
include liver phosphorylase with glucose-6-phos-
phatase (Lowe et al., 1962) and liver phosphorylase
with liver glycogen synthetase (Parr, Teree, and
Larner, 1965). From a consideration of the cases
reported up to that time, Hers (1964) concludes
that the association of two enzymatic defects in one
patient or in the same family has not been proved.
However, not all those reported can be dismissed
as due to imperfect laboratory technique. Multi-
enzyme defects could result from a mutation in
an operator gene controlling more than one adjacent
enzyme (Jacob and Monod, 1961). Such a
possibility is less likely if the gene brancher enzyme
is situated on the X chromosome (see below).

Genetics. Since 2 of the 6 recorded cases were
sibs, the condition is inherited, presumably as a
recessive character. Though an autosomal in-
heritance is most likely, the preponderance of
males, 5 out of 6 cases, suggests that an X-linked
one cannot be excluded. Most types of glyco-
gen storage disease are inherited as an autosomal
recessive. Hers (1964) has, however, suggested a
dominant mode of transmission in type VI, liver
phosphorylase deficiency, to account for the
biochemical findings, though Lôhr (1965) believes an
X chromosomal recessive inheritance would better
accord with the results. No evidence of the hetero-
zygote state could be detected by examination of the
leucocytes from both parents.

Summary

A male infant presented at 8 months of age with
persistent vomiting, hepatosplenomegaly, and failure
to gain weight, for the previous 2 months. The
spleen became progressively more enlarged, the
superficial abdominal vessels became prominent, and
ascites developed. Despite dietary restriction of
carbohydrate, he died at the age of 19 months.
The glycogen isolated from a biopsy of the liver was
sparsely soluble in water, gave a blue colour with
iodine solution, and had a mean chain length of 22
glucose units compared with a normal of 11-14.
The exterior and interior chains were also longer than
normal. Amylpectinosis, Type IV glycogen
storage disease, was diagnosed.

In addition to this abnormal form of glycogen, a
glycogen more nearly normal in structure was also
present in the liver. The liver glycogen synthetase
was found to be very low, and this could account
for the relatively low liver glycogen content com-
pared with that found in other glyco-
genoses.

Amylpectinosis may be readily distinguished
from the more common forms of glycogenosis,
glucose-6-phosphatase deficiency, limit dextrinosis,
and liver phosphorylase deficiency, by the early
development of cirrhosis with portal hypertension,
ending in severe liver failure, as well as by the
relatively simple test with iodine. Examination of
leucocytes from the parents revealed no evidence of
the heterozygote state. Though an autosomal in-
heritance is more likely, an X-linked one cannot be
excluded.

We are grateful to Dr. A. D. M. Jackson, under
whose care the patient was admitted to this hospital,
and to Professor W. J. Whelan for his assistance in the
determination of the glycogen structure.

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