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CONGENITAL PORPHYRIA IN PIGS

by

N.T. Clare

Animal Research Station, Department of Agriculture, Wallaceville.

The term congenital porphyria is used to describe an inborn error of metabolism involving the formation and excretion of porphyrin pigments of abnormal types and in abnormal quantities. The most spectacular aspect of the disease is the deep red brown colour of the bones.

The condition is an extremely rare one but has been recognised in human beings since 1874, 86 cases being recorded in the literature up to 1936. On most of these very little data is available, but one man, Matthias Petry, was studied closely for several years by Fischer in Germany, and during his lifetime provided much of the material on which our knowledge of this condition and of the pigments associated with it was built up. Petry's bones and organs still haunt the literature of the subject.

In animals the occurrence of pigmented bones in slaughter houses has been reported from time to time, and generally wrongly referred to as "ochromosis". (Ochromosis is a quite separate condition in which pigmentation is due to melanins, which are probably derived from proteins.) Until 1935, however, the condition had never been recognised in living animals. It was then found by Fourie in South Africa in a herd of pedigree shorthorns and later in three other herds of cattle in that country. Fourie and Rimington were able to make a unique study of both the chemical and the genetical aspects of the disease in these animals. While five references to pigmented bones in swine have been made over the last forty years no living cases appear to have been studied until the discovery of the animals which are to be discussed in this paper.

Before going further it is necessary to give a brief account of the chemical nature and the significance of the porphyrins concerned in congenital porphyria and their relationship to hemoglobin. Briefly, hemoglobin consists of a protein globin attached to a coloured fraction which is an iron compound of protoporphyrin. Protoporphyrin illustrates the basic structure of the porphyrin group - that is 4 pyrrole groups linked together by CH bridges. The attachment of different radicals in varying order around this ring gives rise to the other porphyrins. Thus in coproporphyrin 4 Me and 4 propionic acid radicals are attached as sidechains; in uroporphyrin 4 acetic and 4 propionic acid radicals.

Variation of the order in which these sidechains are arranged around the pyrrole rings gives rise to different types of porphyrins. Where there are two radicals forming the sidechains, as in coproporphyrin and uroporphyrin, 4 compounds are theoretically possible. Actually only two of these are found to occur in nature, and are designated Type I, and Type III. The protoporphyrin which is found in hemoglobin is a Type III porphyrin, and cannot simply be derived from, or give rise to, a Type I porphyrin. This distinction is important when the question of the origin of the porphyrins in congenital porphyria is under discussion.

Regarding the general properties of porphyrins two are very distinctive - their sharply banded absorption spectra when viewed through a spectroscope and their brilliant red fluorescence under ultraviolet light. Physiologically they are active photosensitising agents.

Congenital porphyria, as it is known in human beings and in bovines, is characterised by the excretion, from birth, of large amounts of both coproporphyrin and uroporphyrin in the urine, and

of coproporphyrin alone in the faeces. Normally coproporphyrin is excreted in very small amounts in both urine and faeces. The amount in congenital porphyria, however, may be increased several hundred times. Uroporphyrin, in the adult at any rate, is a quite abnormal product. The Type 1. porphyrins comprise the greater part of these, only very small amounts of the Type III. being excreted. The hemoglobin of at least one case, however, was shown to be the normal isomer, that is, Type III. The presence of these pigments in the urine often imparts to it a marked red, or sometimes, a brownish colour.

Uroporphyrin is readily precipitated in the presence of calcium salts, and consequently is deposited in the bones during calcification, giving the brown or red coloured bones which is the most striking feature of the condition.

Sensitivity to sunlight has been observed in both human and bovine cases. This is almost certainly due to the presence of porphyrins in the skin, since these pigments are well known as photosensitising agents. Before photosensitivity can occur, however, the skin must be free from other pigments such as melanins, which are capable of providing protection from the parts of the solar spectrum which are absorbed by porphyrins.

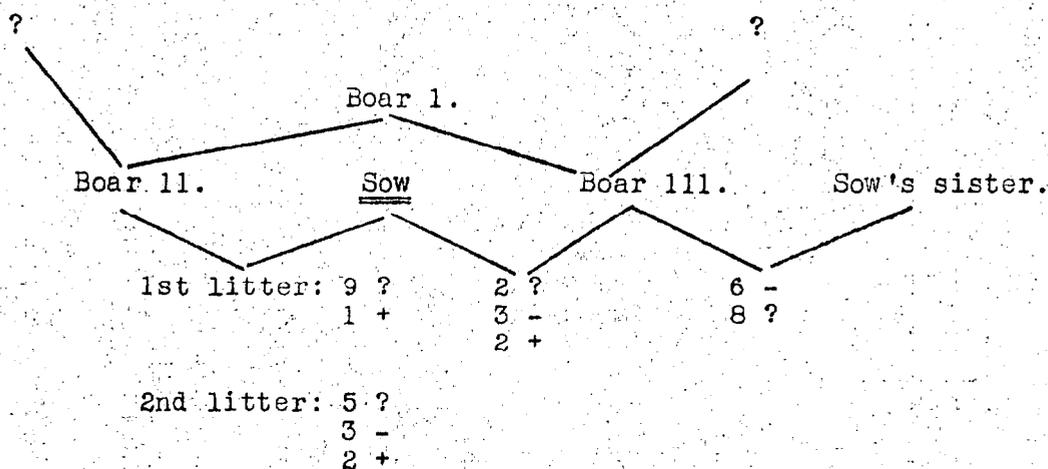
Evidence for the congenital nature of this porphyria is definite from the recorded human cases, although the data available on its occurrence is too incomplete to enable any conclusions on the genetical nature to be made. The South African workers with their herds of pedigree cattle have obtained evidence which leaves little doubt that the condition is inherited as a recessive character. Their evidence for this may be summarised as follows:-

1. Both normal and affected offspring were produced by mating a clinically normal bull to clinically normal cows which are related to the bull by a common ancestry.
2. Experimentally, both normal and affected calves were produced by this bull out of a clinically normal daughter.
3. Calves with congenital porphyria were produced by mating the bull to an affected daughter and by mating one of his sons to one of his daughters.
4. Mating of the original bull to 10 unrelated cows produced only normal offspring (8).

With the foregoing facts in mind, I wish now to report to you the discovery of congenital porphyria in pigs in New Zealand. In January of this year portions of the ribs and sternum of a porker were sent to Wallaceville by Mr J.A. Chenery, Senior Meat Inspector at Waitara, who had observed an unusual pigmentation in them. The bones were dark brown in colour, showed brilliant fluorescence under ultraviolet light, and were found on extraction to contain uroporphyrin and coproporphyrin. The owner of the animal being known, inquiries were made by Mr Stephens, Veterinarian at Stratford, as to its history, and when two further pigs from the same farm were found at the Freezing Works to show this bone pigmentation it was possible to identify the sow which had produced them. Mr Stephens examined this sow and obtained samples of faeces and urine in which considerable quantities of Type 1. porphyrins were detected. These facts, together with the information on the genetical aspect, which I will deal with later, leave no doubt that we are dealing here with a true case of congenital porphyria. This sow is the first living case of the condition in pigs which has been reported.

When first seen by Mr Stephens, the sow had produced two litters which were known to have included pigs with pigmented bones, both litters by the same boar. She was then in pig to a second boar which is a half-brother, on the sire's side, to the first. She has since produced this litter, of which five were dead when

found and two living. The dead ones were examined in the laboratory, two being definite cases of porphyria, while the bones of the other three all showed a slight pigmentation and red fluorescence. However, it is known that the foetal bones of many mammals do contain small amounts of uroporphyrin and I have seen similar slight pigmentation and fluorescence in both rats and rabbits. Whether or not these three slightly pigmented piglets are normal cannot therefore be decided until a number of normal pigs' foetuses have been examined. It is nevertheless of considerable interest that the sow should have produced pigmented litters by two closely related boars. Further information on the breeding of these animals is being sought and arrangements are being made to buy the sow and one of the boars for further study at Wallaceville. The information so far available on the genetics of this condition is summarised in the following diagram. The sow is from a Devon Berkshire cross by a pedigree Tamworth. The boars to which she has been mated are Tamworths.



The animals on which no information is available are marked with an interrogation mark. Some of these are still alive. Animals with pigmented bones are marked +.

A study of the nature and type of the porphyrins excreted aids in distinguishing congenital porphyria from two similar conditions, acute porphyria and chronic porphyria. The acute porphyria, which also appears to be familial, is marked by acute abdominal and nervous symptoms and by the preponderance of porphyrins of Type 111. There is no pigmentation of the bones. Chronic porphyria is a less clearly defined condition in which sometimes the Type 1 isomers preponderate and sometimes the Type 111. Uroporphyrin is frequently not found.

Acute and chronic porphyrias have not been reported in animals. The chemical findings so far in the cases at present under review are summarised in the next table, and compared with composite data on human material, and with the porphyrins obtained by Rimington from bovines.

Congenital Porphyria - Chemical Findings

	<u>Pigs</u>	<u>Man</u>	<u>Cow</u>
Bones	Uro 1 and 111	Uro 1 and 111	Uro 1 and 111
Marrow	Copro	Copro	Uro 1; Copro
Urine	Uro 1 and 111 Copro 1 and 111	Uro 1 and 111 Copro 1	Uro 1 and 111 Copro 1 and 111
Faeces	Copro 1 Proto	Copro 1 and 111 -	Copro 1 Uro Cu Salt
Blood	Copro 1; Uro	Copro	Uro 1; Copro 1

In all these cases the amount of Type 111. porphyrins present represented only a small fraction of the total porphyrins. The nature of the porphyrins present in the pigs establish further the similarity of this condition with congenital porphyria in human beings and in cattle. There are no indications that photosensitivity has occurred in any of the affected pigs. This is not surprising since the skin colouring of both Berkshires and Tamworths is likely to absorb the blue and green rays which are most active in porphyrin photosensitisation.

I mentioned earlier that a great deal of our knowledge of the structure and formation of blood pigments had been obtained by the study of human congenital porphyria. Concerning the actual origin of the pigments little is known. The point on which we can be most definite is that the major part of the abnormal porphyrins is not derived from blood break-down products, since that would involve a transformation from Type 111. to Type 1. isomers. The evidence is against such a change occurring in vivo. Evidence is accumulating, however, that the formation of Type 1. porphyrins occurs simultaneously with the synthesis of the much greater amounts of Type 111. which appear finally as hemoglobin, and their presence in porphyria indicates a derangement, at an early stage, of this synthetic process.

It is hoped that the material available from cases of congenital porphyria in pigs will provide further opportunity for studying these aspects of the problem of pigment formation.

DISCUSSION

Professor C.P. McMeekan:

Commented on possible relationship between porphyria and meat quality in that the abnormality was apparently associated with haemoglobin with which the colour of meat was also linked. He asked whether the carcasses of the affected animals showed any abnormal muscle coloration, especially since dark muscle colour had been reported in respect to affected cattle in South Africa.

Reply:

The muscle of the foetal animals did not show unusual pigmentation, but there were dark areas on some of the organs. Porphyrins were extracted from the flesh, but I don't think that the occurrence of small amounts of porphyrins in flesh is likely to influence the flavour.

Dr F.W. Dry:

On the data presented we may be confident that the condition is inherited in simple Mendelian fashion, and the chances are that the factor is dominant, not recessive.

The chemistry underlying the phenomenon is likely to throw light on the mode of working of the gene concerned, and so to be a contribution in a field in which not much has been done.

Mr R.E.R.Grimmett:

Asked whether porphyrins occurred in plants.

Reply:

The occurrence of coproporphyrin and protoporphyrin in the young leaves of many vegetables and also in yeast has been reported.

Mr A.H. Ward:

Pointed out that the typical case of porphyria in cattle quoted by Clare indicated that the condition was recessive in character in view of the fact that the trait was transmitted from clinically normal parents. On the other hand,