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Short Communication

Inferior dental nerve symptomatology in dental practice needs exclusion of sickle cell disease

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Definition

As this is the most important part of the article, considerable detail is given at length

Sickle cell disease is defined as *two* abnormal beta-globin genes at least one of which is the sickle cell gene denoted by the capital letter S [1]. Other Abnormal beta-globin-genes are denoted either by capital letters of the alphabet C D E G K ... [2] or by the name of the place where the abnormal haemoglobin was found, for example, Haemoglobins Korle Bu [3], Osu-Christiansborg [4], K Woolwich [5] all of which are qualitative beta-globin gene abnormalities. There can, however, be a quantitative beta-globin gene abnormality which then is called beta-thalassemia and which I denote as beta-thal.

Therefore, the phenotypes that fit the definition of sickle cell disease as stated above "*two* abnormal beta-globin genes at least one of which is the sickle cell gene" are SS, SC, SD, SE, SG, SK, SKorle Bu [3], SOsu-Christiansborg [4], SK Woolwich [5], Sbeta-Thalassaemia [6], and so on.

The SS phenotype fits the sickle cell disease definition, but it is the only phenotype that the term Sickle Cell Anaemia is especially devoted to [1,2,5,7].

CAUTION: As Normal beta-globin haemoglobin is denoted with the capital letter A, when this Normal gene A is inherited with any of the Abnormal Haemoglobins the resulting phenotype must never be called a disease, but a Trait. Phenotype "AS" is Sickle Cell Trait, not Sickle Cell Disease [1,2,6-8].

Read this again, please, because the greatest confusion that doctors and the media make today in Haematology is to call Sickle Cell Trait Sickle Cell Disease, and Sickle Cell Disease Sickle Cell Trait [9-11].

Sickle cell crisis: The vital difference is important because it is only Sickle Cell Disease (two abnormal genes) that manifests what is called "sickle cell crisis" when under *in vivo* conditions like dehydration from diarrhea, fever, infections, sepsis, exercise, swimming, trauma, flying in under-pressurized aircraft, cold rainy weather, shock, poor oxygenation the red cells of a person with a sickle cell disease phenotype turn from round to sickle shape while those with sickle cell trait AS behave exactly as persons without any abnormal haemoglobin AA and do not sickle erythrocytes [1,6].

Sickling of red cells in Sickle Cell Traits AS occurs only *in vitro* when a drop of a reducing agent like 2% sodium metabisulphite solution is added to the blood specimen [12,13].

Fraudulent Science has been used to call sickle cell disease people sickle cell traits to confuse phenotypes and that fraud has been used by insurance companies to defraud true sickle cell traits who are entirely healthy. It is dangerous to expose such commercial fraud that was why I was once given 4 bodyguards in Philadelphia [14] at an invited lecture in the presence of Nobel Laureate Linus Pauling who had discovered the molecular pathology of the haemoglobin S gene [15].

While the Media including such important broadcasters as BBC, ITV, and CNN often use the term "sickle cell" to apply

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to both those with Sickle Cell Trait and Sickle Cell Disease, diligent Dental Surgeons and Doctors must never be heard to say to Nurses assisting them "My next patient is Sickle Cell". Nor was it correct for the main news item recently in London to announce: "Body of the missing 18-year-old African student with Sickle Cell has been found". Without phenotype qualification, just to describe a person with the S gene as "Sickle Cell" is not only intolerable but dangerously unfair.

My Philadelphia Lecture entitled "The Vital Difference between Sickle Cell Disease and Sickle Cell Trait" mentioned how a Black American Sickle Cell Trait man (AS) ran at Olympic Games, Mexico City 7,200 ft above sea level where Oxygen concentration is expected to be thinner than at sea level and managed to beat the whole world [16]. He later wanted to buy a house in New York (sea level) only to be told that his insurance would be 150% the normal rate because he had "Sickle Cell Disease".

But what has all this got to do with dental practice?

Answer: When any of the above-mentioned circumstances precipitate a sickle cell crisis from in vivo sickled cells blocking tiny vessels supplying tissues, bone, joints, organs like spleen, liver, skin, placenta, eyes, brain, adrenals, spinal cord, and nerves the symptoms and signs in the patient can be numerous and bizarre.

Because I am the Former Director of the Sickle Cell Clinic for the largest number of patients in the world at the Korle Bu Teaching Hospital in Accra I was able to describe usual and unusual signs and symptoms manifesting themselves in more than 1,500 consecutive patients [1]. One very important symptomatology is what I need to draw Dental Practitioners' attention to.

Fifty years ago, exactly, I described in Lancet [17] a new physical sign in Clinical Medicine that had fooled not a few brilliant Clinicians even in university teaching hospitals. When sickle cell crisis is severe several bones including the mandible are affected. The inferior dental nerve gives rise to 4 nerves one of which is the mental nerve which during its passage through the mental canal on the medial aspect of the mandible emerges at the mental foramen and becomes infarcted during sickle cell crisis.

The Physical Sign I was first to describe in 1972 was a post-sickle cell crisis persistent burning sensation over the chin, spreading to the inside of the lower lip. The area of paraesthesia shrinks over weeks, and by 3 to 4 months becomes a patch of numbness from what I called "peripheral neuropathy of the mental nerve" resulting from infarction of the nerve during the severe sickle cell crisis – a phenomenon that other clinicians later recognized in 1979, 1980, 1982 and 1984 [18-21].

As some authors just observed it was "first described in Africa" without specifying who first published it where and when, I made sure to codify it in The Lancet as the "KANUMBLLL Sign" [22]. The Konotey-Ahulu Numb Lower Lip Lancet Sign.

Relevance to dentistry

1. Do not think this Kanumblll sign is related to previously given local anesthesia

2. As Sickle Cell Disease is found commonly in Greeks, Turks [23], Middle Easters, Asians, and their offspring abroad Sickling Test and quantitative Haemoglobin Electrophoresis need doing to diagnose haemoglobin phenotype not just for Black patients. Note that Sickle Cell beta-Thalassaemia on electrophoresis shows A and S bands with S always greater than A, (written "SA") while in Sickle Cell Trait the Normal Haemoglobin A is always greater than S (written "AS") [1 ,24]

3. Exclude sickle cell disease phenotype before general anesthesia on the patient.

4. Patients with Kanumblll Sign will frequently first go to the Dentist for help, without associating it to the sickle cell crisis they have just recovered from.

5. Identify sickle cell disease patients otherwise accidents could occur in Dental Surgery. Do not use the Sickling Colour Test for diagnosis because it cannot distinguish Sickle Cell Haemoglobin C Disease (SC) from Sickle Cell Trait (AS)!

6. When SC is written in Capitals it means Haemoglobins S and C together comprising Sickle Cell Haemoglobin C Disease (two abnormal haemoglobins) and the SC must not be interpreted as S for the sickle, and C for cell [13].

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