

DOI: 10.36648/1989-5216.12.5.325

Congenital Adrenal Hyperplasia and the COVID-19 Pandemic: “The Calm before the Storm”

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Received date: July 08, 2020; **Accepted date:** July 20, 2020; **Published date:** July 27, 2020**Citation:** Al-Jabri A (2020) Congenital Adrenal Hyperplasia and the COVID-19 Pandemic: “The Calm before the Storm”. Arch Med Vol. 12 Iss.5: 24**Copyright:** ©2020 Al-Jabri A. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Congenital Adrenal Hyperplasia (CAH) is considered one of the most frequent autosomal recessive disorders in humans. It is a quite higher incidence in Saudi Arabia than other countries, estimated by one in 5,000 live births to one in 6400. This disease treated with lifelong cortisone (different types) and recommends increasing the hydrocortisone dose, according to the general sick day rules. The new recommended medication to treat COVID-19 patients is Dexamethasone, which is the most potent form of cortisone and it is glucocorticoid effect higher than hydrocortisone by 25 times. Till the date of write this paper, there is no published case of CAH affected with COVID-19 infection, the reason behind it is not clear but could because they are already on cortisone management.

Keywords: COVID-19; CAH Dexamethasone

To the Editor

There are around 6.5 million newborn screenings worldwide indicate that classical Congenital Adrenal Hyperplasia (CAH) occurs in 1:13,000 to 1:15,000 live births. It is estimated that 75% of patients have the salt-wasting phenotype, the most frequent autosomal recessive disorder in humans [1].

The estimated prevalence worldwide is variable (1 in 10,000) and the annual incidence shows a marked geographical variation from 1 in 409 (Yupik Eskimos) to 1 in 67,000 live births in North America [2,3].

Eighty-two children with CAH were seen at King Khalid University Hospital (KKUH) over 10 years. Of these, 74 (90.2%) were Saudis, and eight (9.8%) non-Saudis. Fifty-nine patients (72%) were 21-hydroxylase deficient. The rest of the patients had either variable degrees of salt depletion or other forms of CAH rather than 21-hydroxylase deficiency [4].

Interestingly, all our patients with both classic and non-classic CAH were homozygous, and this may expect as consanguinity reaches up to 56% in certain parts of Saudi Arabia, this is in contrast to other populations, where the majority of individuals with 21-hydroxylase deficiency CAH are compound heterozygote [5].

Alfadhel and colleague estimate the disorder of CAH in Saudia Arabia of 1:7908 [6], however, their result is not going along with the results reported by Al-Jurayyan et al. which showed that the incidence of CAH in Saudi Arabia was one in 5,000 live birth [4] to one in 6400 [5].

Studies have reported that individuals with adrenal insufficiency have an increased rate of respiratory infection-related deaths, possibly due to impaired immune function and they are more likely to develop a severe course of the disease as well [7,8].

European Society for Pediatric Endocrinology (ESPE) recommends increasing the hydrocortisone dose, according to the general “sick day rules” in children with congenital adrenal hyperplasia.

The suggested oral stress dose for an adult is 20 mg hydrocortisone every 6 hours, if they deteriorate more during acute coronavirus disease 2019 (COVID-19) infection, they advise immediate (self-)injection of 100 mg hydrocortisone intramuscularly, followed by intravenous infusion of 200 mg hydrocortisone per day, or until this can be established, administration of 50 mg hydrocortisone every 6 hours. Also, advise on doses for infants and children [8].

Up to the time of writing this letter, there is no documentation about any patient with congenital adrenal hyperplasia diagnosed to be positive with COVID-19 reported. This absence could be because they are on a good dose of hydrocortisone /prednisolone, and most of the patients and their parents have known about the stress dose of cortisone during the illness as well as during the current COVID-19 pandemic.

In China, the origin of COVID-19, the incidence of congenital adrenal hyperplasia frequency of 1 in 28,000 in the Chinese

population [9]. I am anticipating that a report outlining the association between CAH and COVID-19 may come from countries in which CAH is very common such as our country.

In Saudi Arabia, patients with immunocompromised or who have certain chronic conditions are included in the list of diseases requiring additional precautionary measures to reduce risk of COVID-19 and those patients with CAH known to have an element of immune deficiency because of defective action of neutrophils and natural killer cells [10], as well as having cortisol deficiency and on lifelong treatment, because of this factors, the risk of infection in that patient higher than the normal population by two to eight-fold [11].

The good news is that the Dexamethasone, which is one of the therapeutic agents to treat patients with CAH, has been proposed as a treatment for COVID-19 and clinical trials have been started evaluating this proposal.

Mahase publishes in her clinical trial about using of Dexamethasone to treat COVID-19 patients, she found that low dose dexamethasone reduces deaths in patients hospitalized with COVID-19 who need ventilation by one third (from 41% to 27% with $p=0.00030$) and from 25% to 20% among those needing oxygen [12].

On the other hand, Veronese and colleagues mentioned in their study 201 participants with different stages of pneumonia due to COVID-19, the administration of methylprednisolone significantly reduced the risk of death by 62% [13].

Dexamethasone is a synthetic adrenal corticosteroid with potent anti-inflammatory properties. In addition to binding to specific nuclear steroid receptors, dexamethasone also interferes with NF- κ B activation and apoptotic pathways. This agent lacks the salt-retaining properties of other related adrenal hormones. It is more potent as a glucocorticoid effect than hydrocortisone by 25 times [14] and it's considered as a long-acting glucocorticoid type that we can treat CAH individuals with [15].

Conclusion

I believe that since the Dexamethasone considered as one of therapeutic option to treat severely ill COVID-19 patients, it will help to protect patients with CAH from this serious infection.

Declaration of Competing Interest

The author declares that she has no known competing personal relationships or financial interests that could have appeared to influence the work reported in this paper.

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