## There eb. dystrophic epidermolysis bullosathis type of eb



There are many diseases and genetic mutations in the world, and one of them is epidermolysis bullosa or EB for short. EB is a mainly inherited disease and is a connective tissue disease which causes blisters on the skin and mucosal membranes. Children who have this disease are often nicknamed 'butterfly children', 'cotton wool babies' or 'crystal skin children' in South America. The public first started noticing the disease in the UK in 2004. A documentary was released on 'Channel 4' (a British public-service television broadcaster) about 'the boys whose skin fell off'. The documentary was about the life of Jonny Kennedy, an Englishman that had EB.

There are over 300 mutations that have been identified for this condition. These mutations can range from mild to lethal. These are the types of EB: Epidermolysis bullosa simplexThis is a form of EB that causes blisters on parts of the body that are rubbed a lot. This mutation usually affects the hands and feet of the person with the condition. It is usually inherited in an autosomal dominant way. This affects the keratin (main material that makes up hair, horns, claws, and hooves) genes KRT5 and KRT14. Junctional epidermolysis bullosa Junctional epidermolysis bullosa is an inherited disease (inherited in an autosomal recessive fashion) which affects your laminin and collagen and it is characterized by blistering around the basement membrane.

This mutation also gives the diagnosed person blisters on the feet and hands. It can occur in adults or children, but there is a higher percentage of it happening for children. And out of everyone in the world, less than one person out of a million are said to have this form of EB.

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Dystrophic epidermolysis bullosaThis type of EB affects the skin and other organs in your body. Dystrophic epidermolysis bullosa is caused by a genetic defect or mutation in the type of human COL7A1 gene. DEB can either be autosomal recessive or dominant. It is also the most severe form of EB out of all the forms.

Epidermolysis bullosa acquisitaAcral peelingThe three main types of EB are EB simplex (EBS), junctional EB (JEB), and dystrophic EB (DEB). One of the ways to treat EB is by gene therapy. The idea of gene therapy was published on November 1st in JAMA (Journal of the American Medical Association). Gene therapy itself tries to treat genetic diseases by correcting what is wrong with the gene. It helps the patients with EB by grafting sheets of the patient's healthy skin and putting it on the open wounds on their body caused by the disease. Gene therapy works by locating a reliable system to carry the right gene to the cells that are affected by EB. The gene needs to be delivered to the cells and work correctly without causing bad effects on the patient's body.

When they were studying gene therapy, 4 adults with DEB (dystrophic epidermolysis bullosa) were given the skin grafts for their wounds. In the phase-one trial that the dermatologists and scientists did, the wounds healed amazingly. For every cure or product in the world, there are always advantages and disadvantages, and these are the ones for gene therapy. The advantages are: Gene therapy has the potential to heal and cure some diseases and disorders for those who are diagnosed with a disease/disorder, and it can stop the disease from taking hold in people who have the possibility of getting a disease. Another advantage is, if you were to do gene https://assignbuster.com/there-eb-dystrophic-epidermolysis-bullosathis-type-of-eb/

therapy on the reproductive cells of a patient with EB or another skin disease, you would be able to stop the offsprings from inheriting a disease or disorder from their parents. Now to the disadvantages; One of the main disadvantages of gene therapy is that it is extremely expensive. Gene therapy in Europe in 2012 cost around \$1 million, which is a lot of money that not everyone can afford.

Secondly, if scientists were to find the gene that caused you to look youthful and beautiful, the cosmetic and beauty industry could use it to make people more 'beautiful'. This is not what gene therapy is supposed to be used for, and since there are much more important diseases to cure than looking beautiful, this is not a good thing that should happen. However, if this does happen, then the government will have to decide whether fixing a big nose, or a wrinkly forehead will be as important as curing genetic diseases such as EB.