

**IT'S IN THEIR GENES!**

While there are different forms of progeria, the classic type is Hutchinson-Gilford Progeria Syndrome (HGPS), which was named after the doctors who first described it in England—Dr Jonathan Hutchinson in 1886 and Dr Hastings Gilford in 1897. Its extreme rarity and sporadic occurrence were posing a challenge to researchers, who were trying to identify its underlying genetic cause. However, recent studies show that HGPS is caused by a mutation in the gene called LMNA. According to scientists at the Progeria Research Foundation in the US, the LMNA gene produces the lamin A protein, which is the structural scaffolding that holds the nucleus of a cell together.

Researchers now believe that the defective lamin A protein makes the nucleus unstable. That cellular instability leads to premature ageing. Although they appear healthy when born, children with progeria begin to display many characteristics of accelerated ageing when they are 18-24 months of age. The symptoms include loss of body fat and hair, skin that looks aged, stiffness of joints, hip dislocation, impaired or absent sexual maturation, generalised atherosclerosis (heart disease) and stroke. Current research shows that children with progeria die of atherosclerosis at an average age of 13 years.

which has been looking after the Khans and working in collaboration with the University of Basel for the last six years.

"There may be no cure for the condition, but over the last couple of years we have seen that a healthy diet and lifestyle changes can enhance the life expectancy and its quality," says Chattopadhyay. Traditionally, not many progeria victims live beyond their early 20s. "We have been able to maintain the children's weight and ensure that the loss of bone-joint width and skin-fold thickness is minimal," says Dr Ajay Mukherjee, physiologist and senior researcher with the project.

But, life is far from normal for the boys. Rashes and boils are a recurring problem, while the total absence of fat makes their bodies susceptible to frequent injuries. "They deal with regular problems associated with old age such as osteoporosis, arthritis and pigmentation of the skin. Moreover, they are so fragile that if the fan is in full speed, it is difficult for them to maintain their balance. And they need help for even simple activities like changing clothes," says Chattopadhyay.



EXCLUSIVE

# Twinkle in wrinkle

Despite advances in medicine, India's only 'progeria family' continues to suffer

By Jisha Krishnan

**T**he two pairs of protruding alien-like eyes unnerve me. One of the boys laughs, revealing a second row of unaligned teeth. Somewhere in the background, I barely hear the boys' mother, Razia Khan, speak of the nightmarish life the family was forced to lead at their village in Chapra, Bihar: "The children were called all sorts of names—*bhoot* (ghost), *shaitan* (devil) and what not! Nobody wanted to talk to them. They used to sit locked up within the house all day, no school, no play, nothing."

They are unique. They are scary. And, at that point in time, just too close for comfort. Flanked by Ikramul Khan, 22, and his brother Ali Hussein, 11, in their Kolkata residence, I struggle not to make my discomfort obvious. What I need is some distraction, I decide. But there's just too much to ignore: their almost translucent skin, frail bodies with not an iota of fat, thinning hair, weird-looking fingers...

After what seems like an eternity, I muster enough courage to start a polite conversation. Ali is vivacious. In no time, we exchange views about the importance of a good pair of denims in one's wardrobe. "I don't quite fancy these formal trousers," he says, referring to what he is wearing. He sounds more like the enthusiastic 11-year-old that he is than the sexagenarian in whose body he has been trapped.

But then, there's no agility in his

movements, rather a marked caution in the way he sits in the chair or moves slowly across the corridor. His bones are too brittle and joints too stiff, severely impairing his movements. Ikramul is reclusive. He speaks only when spoken to, mostly in monosyllables. He is yet to recover from a depression he fell into after the death of his sister Rehana last year. "They were of the same age group and were really close. He probably knows that he is next," says Razia.

Ikramul keeps dabbing his eyes frequently. "Is he crying?" I ask. "No, it's just that his eyes keep watering because of his age," explains Razia. According to medical reports, Ikramul's body is like that of an 85-year-old's. Rehana was 23 when she died a 'natural death' last year and her physical condition was that of a 90-year-old.

Bisul Khan, 45, and his wife, Razia, 40, had seven children in all. Five of them were born with progeria, which is characterised by premature ageing. One died prematurely and another died of pneumonia at the age of 16. Their daughter Sanjeeda, 20, is a progeria carrier and their youngest child, Chanda, is healthy.

Though the first cases of progeria, which means prematurely old in Greek, were identified more than a century ago, only 48 cases have been reported worldwide. More people will come to know of progeria with



All Hussein	Normal
Age: 11 (medically, 60 plus)	
Weight: 12kg	25-30kg
Height: 116.9cm	134-138cm
Arms: 12.7cm	16-20cm
Chest: 42.4cm	45-47cm

(Estimates of the vital stats of regular individuals were arrived at on the basis on interactions with experts.)

the release of the Amitabh Bachchan-starrer *Paar* in which he plays the role of a progeria patient. Interestingly, Abhishek Bachchan and Vidya Balan play the role of Amitabh's parents.

The Khans are the only family in India where more than one member suffers from progeria. Until about six years ago, the family had no clue about their medical condition. "We took the children to a lot of doctors—from child specialists to skin specialists and homoeopathy practitioners.



**Braving odds:** Bisul and Razia with their children Ali (left), Chanda and Ikramul

Ikramul	Normal
Age: 22 (medically, 85 plus)	
Weight: 13.5kg	55-60kg
Height: 123cm	165-175cm
Arms: 11.5cm	24-26cm
Chest: 46cm	80-95cm

But no one knew what the problem was. If someone had explained the condition to us, we wouldn't have had so many children," says Bisul, who works as a security guard in Kolkata. On more than one occasion, the family contemplated suicide.

It was a chance meeting with paediatrician Chandan Chattopadhyay at the Institute of Child Health in Kolkata in 2002 that made the Khans aware of progeria. Until then, researchers had believed that the

condition was non-hereditary. But a two-year study of the family proved that progeria was actually an inherited and recessive condition. About two years ago, the UK's Channel 4 aired a documentary on the family entitled, *Bodyshock: The 80-year-old children*.

A team led by Dr Chattopadhyay at the University of Basel, Switzerland, has been constantly monitoring the progress of the family. Besides understanding the intricacies of the condition, researchers are hopeful

that by studying the ageing process in this unusual form they may get new insights into how ordinary human beings grow old.

"These are children of average intelligence, in whom the ageing process is normal up to the age of seven. After that, it's a downhill journey. Life zooms past in a fast forward mode and soon they find themselves grappling with old age," says Sekhar Chattopadhyay, general secretary of S.B. Devi Charity Home, Kolkata,

# I didn't recognise dad!

By **Abhishek Bachchan**

I hadn't heard of progeria until about two years ago, when Balki [director R. Balakrishnan] first told me about the film [*Paa*]. I was intrigued by the rare genetic condition. Once I started reading about its effect on the human body and saw pictures of patients, I was quite taken aback. That's the effect it would have on anyone!

To see my dad play the role of a person suffering from the condition was an unusual experience. On the first day, when we had the look test and dad came out in his make-up, I didn't recognise him. It took me 10 minutes to realise that he was the man standing right next to me, and I had been looking for him all around. That was weird!

But playing dad to him wasn't that weird. It was quite easy actually.

Psychologically, they have demons to fight. Deprived of a social life, Ali and Ikramul spend most of their time watching TV or downloading songs on the mobile phone. Two years ago, the family was invited to a conference in San Francisco, but the children refused to go. "What if they throw us down from the plane?" they asked their parents. The trip didn't happen, and much of the fear persists.

The Khans look upon Chattopadhyay as a family member. "He looks after us, bears all our expenses, provides us accommo-

Maybe because the dad-son relationship is more of friendship in the film. Besides, at the end of the day, we are just actors doing a job.

Though Auro, the 12-year-old character in the film, is shown to be suffering from the condition, the film isn't really about progeria. We haven't delved into the many hardships that these unfortunate children have to live through; rather we have tried to show a happy, active child, who has a zest for life. Auro is a fun character, without being frivolous.

We have made a conscious effort not to use progeria as a marketing point. We are aware of the 'progeria family' based in Kolkata; we have seen the documentary on them, too. Once the film releases [on December 4], many more people will be aware of the condition. And, that's a good thing. But ours is only a film, which takes necessary cinematic liberties in the portrayal of the character.

was something that the children eagerly looked forward to. "There hadn't been a single celebration in our family before. By the grace of God, we managed to get a well-employed, good-looking man for our daughter," says Razia. Sanjeeda, though a carrier, is pregnant with a healthy baby, due for delivery this month.

Ali has an idea: "Why not get him married?" Ikramul can't resist a smile. But he isn't interested in marriage. He has one wish, though. "Didi, can you get us to meet Salman Khan?" he asks. "And, Shah Rukh Khan, too?"