



In a remote central Bihar village where life is as drab as cowdung cake and stimulation emanates only from an earthen cup of country liquor and a stretched puff of Murshidabad bidi, the Hutchinson-Gilford Progeria Syndrome (HGPS) should sound as an impossible Latin tongue twister. However, with sudden materialisation of six "satanic siblings," HGPS is as familiar as the heat and dust of the countryside.

This weird disease gave them the scare of their life about a decade ago, only to afford them a sense of identity today. This village (name withheld) is known in Europe for housing a Progeria family, the first of its kind in the world.

"Bizarre" because their existence scared the naive villagers, who mercilessly ostracised them, calling their progenies children of the "Jinn" whose presence could signal apocalypse.

This is a story of poor Bisul Khan and Razia and their children Ravina, Rehana, Ikramul, Ali, Sangita and Gulavsa. Two others were consumed by the disease that eventually twisted the family's entire destiny forever.

Subsequently, the strange and rare syndrome felled Ravina and Rehana as well.

While the Almighty has spared Gulavsa and Sangita, brothers Ikramul and Ali are awaiting their turn. They have been sailing in the same boat as Auro of the much-talked film *Paa*, starring Amitabh Bachchan as a progeria victim.

The real life story would have never come to life had Bisul Khan, a part-time security guard, and wife Razia not chosen to travel all the way to Kolkata 650 km away, where Dr Chandan Chatterjee bumped into them.

Brother Ikramul and sister Rehana were in a worse condition as they thinned fast, their skins crumpled beyond imagination, eyes sunk into the cavity giving them a very old look. "Villagers called them creation of shaitans and did not allow them into their houses," said Bisul.

Dr Chatterjee, who had done field work on Progeria, referred them to the S Devi Charity Home which is involved in research works and charity in many other projects with Swiss collaboration.

"You can call it an untimely ageing of an individual. In this a child grows normally till 6 or 7 years, after which in one year the age of his appearance grows by 45 years. This rate of ageing continues and

normally such children do not live beyond their 20s. Their early death can be delayed but not avoided," said Dr Shekhar Chatterjee, general secretary, Charity Home.

"These people have been like angels to us after our villagers pushed us to the brink of suicide," said Razia. She recalls how one day she had strayed into a marriage

ceremony only to be hounded away "because they thought that my daughters were ghosts and they were bad omen on any family." This was followed by some villagers taking recourse to black magic against the family. Soon her daughter Ravina passed away only stoking the ghost gossip.

"We thought, in any case none of our children was going

to survive, so we decided to end our lives en masse," reminisces Bisul. But somehow they gathered courage and gave life a last chance by visiting Kolkata "where we found these people who are more than our relatives, sustaining, treating and maintaining all of us. They have even constructed a *pusca* house for us in Bihar."

Dr Chatterjee said: "This is a rare genetic problem. Till recently there were about 48 persons in the world suffering from this disease." Out of them three died and now there are 45. People suffering from Progeria are in South Africa, Australia and the UK.

"But this family in India is unique because it has helped researchers go a long way in proving that the disease has a genetic core. In fact the family, the first of its kind, is called the Progeria family world over," the doctor said, adding the disease can occur in one out of 80-90 lakh people.

Progeria is a genetic condition that causes a new mutation and is not usually inherited, "although there is a uniquely inheritable form." This is different from another rare, but similar premature ageing syndrome — Dyskeratosis Congenita — which is inheritable and can occur



World's first 'Progeria Family' was humiliated by neighbours and shooed away as ghosts. The family is now living with an NGO and will be taken to Europe soon for further research on them. Out of eight children, four are already dead and of the remaining four, two are normal, but they are still the carriers

pioneer

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Food problems are... The only superpower in... is climbing the... on the no-food index... the administrators sleepless... and hard-work days

Factoids | II

On the confluence of the Ganga and the Yamuna, five km from Badrinath, there lies a not so well known temple of Hanuman where most pilgrims take a stopover. A report on this heritage spot



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The basic human sense of smell is not just confined to living beings. Scientists across the world are using this human technology for further scientific development. A report

BODY SHOCK

She was 23 but looked 80. He is 21, but shows signs of extreme old age. Their skin is peeling off and bones are as fragile as an old person's. The disease they suffer from was brought into sharp focus with Amitabh Bachchan's

latest flick *Paa*. But the real life family of Progeria children in Kolkata has been bearing the burden of this rarest of rare ailment for more than three decades. **SAUGAR SENGUPTA** meets the family to tell you more

THE LATE AMBASSADOR



John Tacket (1988 – 2004): The Progeria Research Foundation's First Youth Ambassador

This interview was taken in 2001 when John was 13-and-a-half-years-old. He wanted other kids to know about Progeria, and that he was really no different than other boys his age. John was an amazing young man whose courage and wonderful sense of humour continues to inspire us all

Tell us about school.

I'm in the eighth grade. I go to public school and take the bus every day. It's "loaded" with kids! I kind of like school — it's a lot of work, but I like that my friends are there.

What kinds of activities are you involved in?

I play street hockey, play drums, my dad and I have a knife collection, I belong to the roller hockey and floor hockey clubs, and the academic track club which holds meets for lots of subjects, like Math, Science and drama.

I really want to learn how to ice skate so I can play ice hockey. I also love to draw.

What do you do with your free time?

Homework, playing drums, hanging out with my friends and watching TV. The Disney Channel is my favourite.

Tell us about a typical day for you.

I'm up at 6:30 am, I go to school, then baby-sit my nine-year-old sister after school until my mom gets home from work. I also watch TV and play with friends after school and on weekends.

What is your favorite thing to do?

Play drums, play street hockey and be with my family.

What is your least favourite thing to do?

Get up for school!

How long have you known that you have Progeria?

As long as I can remember.

What do you say to people who ask why you look different?

I tell them I have a disease called Progeria.

What do you do or say to people who stare at you?

That bugs me a lot. I'd rather they come over and say "hi" and ask me directly rather than have them stare from a distance. I think it's rude to stare. Sometimes I wave to the kids, and

they wave back.

What are your biggest challenges physically?

It's hard to play basketball — I need a smaller ball and a shorter hoop which I have at my house so a lot of times my friends come over to play basketball.

Sometimes I get tired, but I just sit down for a couple of minutes and then I'm ready to go. A lot of times it's my friends that want to rest before I do! I have a hard time carrying a lot of books, so I leave five minutes early from my classes and go to my locker after every class to switch books for my next class.

What would you like kids to know about people suffering from Progeria?

We're no different; we do the same things other kids do. Don't be afraid to talk to us.

If you were a parent of a child with Progeria, what would you say to your child as he/she begins to realize that he/she looks different?

I'd say yes, you're different but only on the outside. You can do anything you want and don't let anyone tell you any differently. And I would tell them to treat us the same as any child.

If you were a parent of a child with Progeria, what would you tell your child when he/she discovers that he/she may not live as long as other people?

Well, no one really knows how long they're going to live, so I don't worry about it.

What advice do you have for children with Progeria?

Keep going, follow your dreams. My dream is to be a studio drummer and advertising agent.

Is there anything else you want to say to everyone reading this article?

We're just like any other kids, if we're four, we act like it, and so on with the age. Say "hi" to the next person you see with Progeria or to any kid who looks different.