

感覚や思考能力はそのまま、筋力だけが衰える ALS とは

ALS: 筋萎縮性側索硬化症 (指定難病)

突然発症し短期間のうちに進行する深刻な神経難病。

日本における患者数：約 1 万人
毎年 1,000~2,000 人が新たに診断されている。
発病率：10 万人あたり 1.1~2.5 人
平均寿命：2~5 年
難病のなかでも最も酷な病気といわれている。
極めて進行が速いが、現在、根治療法はない。




どんな人に多い？
男女比：男性が女性に比べて 1.2-1.3 倍
発症年齢：平均 59 歳
最もかかりやすい年齢層は 60~70 歳台

ALS の原因は？ 不明
ALS は遺伝する？多くの場合は遺伝しない



筋肉そのものの病気ではなく、筋肉を動かし、身体を動かすための神経系（運動ニューロン）だけが障害を受け徐々に壊されてしまう病気です。

ALS はどんな症状がおきる？

- 手や指が使いにくい 
- 肘から先の力が弱くなる
- 肩が上がらない 
- 足に力が入らない
- よく転ぶ筋肉が痙攣する
- 話しにくい 
- 食べ物がのみ込みにくい

症状が進むと？

- 手足の麻痺による運動障害
- コミュニケーション障害
- 嚥下障害（食べ物が飲み込めない）
- 呼吸障害

4 つの症状がすべてあらわれるようになる
意識や五感最後まで正常で、知能の働きも変わらない

運動ニューロンは、脳の命令を筋肉に伝える役目をしています。
運動ニューロンが侵されると手や足、顔など、自分の思いどおりにからだを動かす事が出来なくなり、コミュニケーションも困難になります。呼吸筋も次第に弱くなって呼吸が困難になります。
(日本では 7 割の方が人工呼吸器を付けず死を選択している。)

しかし、知覚神経や自律神経は侵されないため、五感（視覚、聴覚、臭覚、味覚、触覚）、記憶、知性を司る神経には原則として障害はみられません。
知性、体の感覚、視力や聴力、内臓機能などはすべて保たれることが普通です。

皮膚をつねられたとき、痛いと感じ、つねられた手をひっこめるでしょう。痛いと感じるのは「知覚神経」、手をひっこめるのは「運動ニューロン」の働きです。ALS になると痛いという感覚はありますが、手をひっこめることができなくなります。

ALSの最終段階、TLSとは？ TLS [Totally Locked-in State (完全な閉じ込め状態)]

全ての筋肉が完全に停止
眼球運動やまばたきもできない

周囲との意思伝達を奪われてしまう

しかし

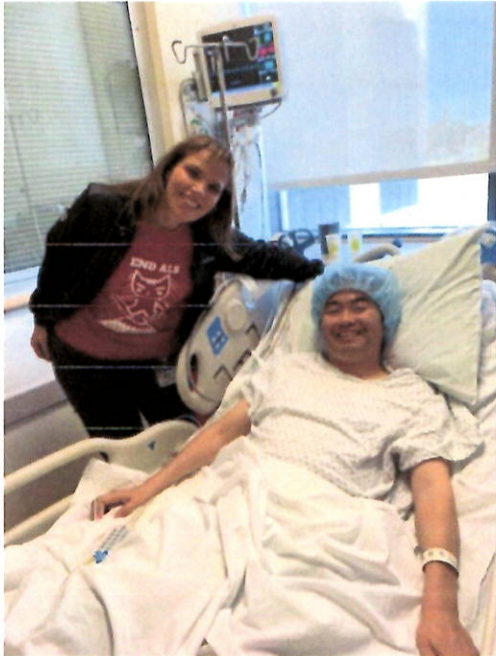
その状態になったとしても なお、意識や五感、知能の働きは正常のまま

Hope for an ALS cure for Squamish ski instructor, and others like him

Local man finishing stem cell trials in California, looking for next option and hope Sea to Sky folks can help

[Jennifer Thuncher](#) / Squamish Chief

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Winnie and Ryoichi Hosoya.
Photo: COURTESY THE HOSOYA FAMILY

In the summer of 2018, Squamish's Ryoichi Hosoya felt a weird feeling in his right foot.

Hosoya, a ski instructor at Whistler Blackcomb for more than two decades, waited it out, but when his foot didn't get better, he sought out physiotherapy.

It was determined he had something called foot drop, or a weak foot, said his wife Winnie Hosoya, who spoke to The Chief on behalf of the couple on June 4.



Ryoichi Hosoya, before ALS.- Courtesy the Hosoya family

A few months of unsuccessful therapy later, and approaching his season on the mountain, he started to seek out other medical diagnoses. Doctors, too, thought at first he had foot drop.

Many tests later, on March 9, 2019, he was diagnosed as having Amyotrophic Lateral Sclerosis (ALS, also known as Lou Gehrig's disease).

"ALS is the most terrible disease. The person who has ALS has just the worst — their body just keeps deteriorating and eventually they will stop breathing and they will just die without even having a voice. It is just horrible, horrible suffering," said Hosoya.

She acknowledges she didn't know anything about the disease before he was diagnosed.

Part of the reason for the couple going public is to raise awareness of the disease in the Sea to Sky.

"People in the Sea to Sky are great, and they just need to know that there is not just my husband.... there are actually quite a few ALS patients in the Sea to Sky," she said.

Once diagnosed, the disease progressed quickly.

"We didn't think he would last to the end of the winter," Hosoya said, reflecting on that dark time. "He was pretty much on 24-hour care, and I had to feed him. I couldn't even touch him because he was in so much pain."

Fast forward — having exhausted his B.C. options, The Hosoyas have been in Anaheim, California for a [Phase 3 stem cell therapy](#) trial since December.

He has had three cell transplants in total, the last one at the end of March

Since then, COVID-19 has swept the globe and more recent protests against police brutality have filled the streets across North America.

"Funny enough... we were going through this by ourselves, but then we feel like, 'Oh Gosh, this isn't so bad. The world is actually in a lot of pain together with us,'" she said. "That is a selfish feeling — but we aren't alone."

And there have been positives along the way.

The trial has shown promise, for example.

"We feel like we have seen improvements. Not major, it is not like he is standing up again and running, like some people claim on the internet," she said. "But we do see the progression slow down."

Hosoya said her husband isn't in pain and currently feels happy.

The **trials** are finishing up in mid-June.

"It is a better result, but is not a cure, unfortunately," she said. Next, they want to try some private company drug trials, but they are costly.

Thus, there is a [Go Fund Me campaign](#) set up by Squamish a neighbour to help the couple access more options.

"We just have to keep looking," Hosoya said, adding her husband wants to help with the trials not just for himself, but so that eventually there will be a cure for ALS.

"We are still hopeful," she said. "We will still keep fighting."

June is ALS Awareness Month in B.C.

More than 400 people in B.C. are living with ALS; over 3,000 in Canada, according to the ALS Society of British Columbia.



The Squamish group, who attended the annual ALS walk in 2019. -
Courtesy the Hosoya family

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How it works

Start a GoFundMe



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Help Ryoichi (Yoyo) fight ALS

\$16,865 raised of \$20,000 goal

105 donors
265 shares
105 followers

Share

TEAM FUNDRAISER



Ko Homma and Winnie Hosoya are organizing this fundraiser on behalf of Winnie Hosoya.

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Medical, Illness & Healing

I've had the pleasure of working with Ryoichi at both Ski School and Lift Ops for Whistler Blackcomb and later became his neighbour in Squamish. It was a shock to me to hear last spring that he had ALS (Lou Gehrig's disease) and then to see firsthand how debilitating ALS is.

Ryoichi ("Yoyo") Hosoya and his wife

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\$100 • 11 d

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