Congenital Mirror Movement Disorder

Subjects: Genetics & Heredity

Contributor: Nicole Yin

Congenital mirror movement disorder is a condition in which intentional movements of one side of the body are mirrored by involuntary movements of the other side.

Keywords: genetic conditions

1. Introduction

For example, when an affected individual makes a first with the right hand, the left hand makes a similar movement. The mirror movements in this disorder primarily involve the upper limbs, especially the hands and fingers. This pattern of movements is present from infancy or early childhood and usually persists throughout life, without other associated signs and symptoms. Intelligence and lifespan are not affected.

People with congenital mirror movement disorder can have some difficulty with certain activities of daily living, particularly with those requiring different movements in each hand, such as typing on a keyboard. They may experience discomfort or pain in the upper limbs during prolonged use of the hands.

The extent of the mirror movements in this disorder can vary, even within the same family. In most cases, the involuntary movements are noticeable but less pronounced than the corresponding voluntary movements. The extent of the movements typically stay the same throughout the lifetime of an affected individual.

Mirror movements can also occur in people who do not have congenital mirror movement disorder. Mild mirror movements are common during the normal development of young children and typically disappear before age 7. They can also develop later in life in people with neurodegenerative disorders such as Parkinson disease. Mirror movements may also be present in certain other conditions with a wider range of signs and symptoms (syndromes).

2. Frequency

Congenital mirror movement disorder is a very rare disorder. Its prevalence is thought to be less than 1 in 1 million. Researchers suggest that some mildly affected individuals may never be diagnosed.

3. Causes

Congenital mirror movement disorder can be caused by mutations in the *DCC* or *RAD51* gene; mutations in these genes account for a total of about 35 percent of cases. Mutations in other genes that have not been identified likely account for other cases of this disorder.

The *DCC* gene provides instructions for making a protein called the netrin-1 receptor, which is involved in the development of the nervous system. This receptor attaches (binds) to a substance called netrin-1, fitting together like a lock and its key. The binding of netrin-1 to its receptor triggers signaling that helps direct the growth of specialized nerve cell extensions called axons, which transmit nerve impulses that signal muscle movement. Normally, signals from each half of the brain control movements on the opposite side of the body. Binding of netrin-1 to its receptor inhibits axons from developing in ways that would carry movement signals from each half of the brain to the same side of the body.

Mutations in the *DCC* gene result in an impaired or missing netrin-1 receptor protein. A shortage of functional netrin-1 receptor protein impairs control of axon growth during nervous system development. As a result, movement signals from each half of the brain are abnormally transmitted to both sides of the body, leading to mirror movements.

The *RAD51* gene provides instructions for making a protein that is also thought to be involved in the development of nervous system functions that control movement, but its role in this development is unclear. Mutations in the *RAD51* gene result in a missing or impaired RAD51 protein, but it is unknown how a shortage of functional RAD51 protein affects nervous system development and leads to the signs and symptoms of congenital mirror movement disorder.

3.1. The Genes Associated with Congenital Mirror Movement Disorder

- DCC
- RAD51

4. Inheritance

In most cases, including those caused by mutations in the *DCC* or *RAD51* gene, this condition is inherited in an autosomal dominant pattern, which means one copy of the altered gene in each cell is sufficient to cause the disorder. In most cases, an affected person has one parent with the altered gene. Some people who have the altered gene never develop the condition, a situation known as reduced penetrance.

Research suggests that in rare cases, this condition may be inherited in an autosomal recessive pattern, which means both copies of the gene in each cell have mutations. The parents of an individual with an autosomal recessive condition each carry one copy of the mutated gene, but they typically do not show signs and symptoms of the condition.

5. Other Names for This Condition

- · bimanual synergia
- · bimanual synkinesis
- CMM
- · congenital mirror movements
- · mirror movements

References

- 1. Depienne C, Bouteiller D, Méneret A, Billot S, Groppa S, Klebe S, Charbonnier-Beaupel F, Corvol JC, Saraiva JP, Brueg gemann N, Bhatia K, CincottaM, Brochard V, Flamand-Roze C, Carpentier W, Meunier S, Marie Y, Gaussen M, Stevani n G, Wehrle R, Vidailhet M, Klein C, Dusart I, Brice A, Roze E. RAD51haploinsufficiency causes congenital mirror move ments in humans. Am J Hum Genet. 2012 Feb 10;90(2):301-7. doi: 10.1016/j.ajhg.2011.12.002.
- 2. Depienne C, Cincotta M, Billot S, Bouteiller D, Groppa S, Brochard V, Flamand C, Hubsch C, Meunier S, Giovannelli F, Klebe S, Corvol JC, Vidailhet M, Brice A, Roze E. A novel DCC mutation and genetic heterogeneity in congenital mirror movements. Neurology. 2011 Jan 18;76(3):260-4. doi: 10.1212/WNL.0b013e318207b1e0.
- 3. Franz EA, Chiaroni-Clarke R, Woodrow S, Glendining KA, Jasoni CL, RobertsonSP, Gardner RJM, Markie D. Congenit al mirror movements: phenotypes associated with DCC and RAD51 mutations. J Neurol Sci. 2015 Apr 15;351(1-2):140-145. doi:10.1016/j.jns.2015.03.006.
- 4. Gallea C, Popa T, Hubsch C, Valabregue R, Brochard V, Kundu P, Schmitt B, Bardinet E, Bertasi E, Flamand-Roze C, Al exandre N, Delmaire C, Méneret A, Depienne C, Poupon C, Hertz-Pannier L, Cincotta M, Vidailhet M, Lehericy S, Meuni er S, Roze E. RAD51 deficiency disrupts the corticospinal lateralization of motor control. Brain. 2013 Nov;136(Pt 11):33 33-46. doi: 10.1093/brain/awt258.
- 5. Galléa C, Popa T, Billot S, Méneret A, Depienne C, Roze E. Congenital mirrormovements: a clue to understanding bim anual motor control. J Neurol. 2011Nov;258(11):1911-9. doi: 10.1007/s00415-011-6107-9.
- 6. Klar AJ. Selective chromatid segregation mechanism invoked for the humancongenital mirror hand movement disorder development by RAD51 mutations: ahypothesis. Int J Biol Sci. 2014 Sep 10;10(9):1018-23. doi: 10.7150/ijbs.9886.
- 7. Méneret A, Depienne C, Riant F, Trouillard O, Bouteiller D, Cincotta M, BitounP, Wickert J, Lagroua I, Westenberger A, Borgheresi A, Doummar D, Romano M, RossiS, Defebvre L, De Meirleir L, Espay AJ, Fiori S, Klebe S, Quélin C,Rudnik -Schöneborn S, Plessis G, Dale RC, Sklower Brooks S, Dziezyc K, Pollak P,Golmard JL, Vidailhet M, Brice A, Roze E. Congenital mirror movements: mutationalanalysis of RAD51 and DCC in 26 cases. Neurology. 2014 Jun 3;82(22):1999 -2002.doi: 10.1212/WNL.00000000000000077.
- Peng J, Charron F. Lateralization of motor control in the human nervoussystem: genetics of mirror movements. Curr Op in Neurobiol. 2013 Feb;23(1):109-18.doi: 10.1016/j.conb.2012.08.007.

9.	Srour M, Rivière JB, Pham JM, Dubé MP, Girard S, Morin S, Dion PA, Asselin G, Rochefort D, Hince P, Diab S, Sharafa
	ddinzadeh N, Chouinard S, Théoret H, CharronF, Rouleau GA. Mutations in DCC cause congenital mirror movements.
	Science. 2010 Apr 30;328(5978):592. doi: 10.1126/science.1186463.

Retrieved from https://encyclopedia.pub/entry/history/show/11290