Certification of a Pilot with Charcot-Marie-Tooth Disease

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BACKGROUND: Charcot-Marie-Tooth disease (CMT) is a rare hereditary motor and sensory neuropathy. This is a report of a pilot with this condition with a discussion of the challenges for the regulator in the assessment for medical certification of pilots with a neurological disability.

CASE REPORT: A pilot with CMTX1 declared his condition to the United Kingdom Civil Aviation Authority when his brother was diagnosed with the same condition. Apart from high arched feet and some difficulty playing sports, he had no problems until his mid-forties, when he very slowly developed increasing weakness with foot dorsiflexion and later wasting and weakness of the small hand muscles. He reported no problems with any flying activity. On clinical examination, it seemed likely that the disability would have an impact on his ability to undertake all the flying tasks of a commercial pilot, including those required in emergencies.

DISCUSSION: A modified Medical Flight Test (MFT) specifically tailored by the regulator to test areas of functional impairment allowed the successful certificatory assessment of a pilot with this condition; an approach which could apply to any pilot with a rare neurological disability.

KEYWORDS: hereditary motor and sensory neuropathy, Charcot-Marie-Tooth, occupational functional assessment.

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Charcot-Marie-Tooth disease (CMT) is a hereditary motor and sensory neuropathy which has multiple variants, now defined by known genetic abnormalities: CMT 1 to 7 and X-linked, each with several subtypes.[7] Over 90% are due to mutations in one of four genes: PMP11, MPZ, GJB1, and MFN2.[4] The mode of inheritance may be dominant or recessive.[6] The average prevalence is about 40/100,000, varying from 10–82/100,000 in different reports.[1] Clinically they all present with a length-dependent peripheral neuropathy and the presentation is similar across the genetic variants. Some subjects present in childhood and others in adult life with a mixed motor and sensory neuropathy, usually with motor predominance, although in CMT2 sensory deficit predominates. CMT1 is a demyelinating neuropathy and CMT2 an axonal neuropathy.[4] Pes cavus is the first sign due to weakness of the small foot muscles and may be the only sign in childhood and adolescence; about 50% of children with pes cavus turn out to have CMT disease.[3] This is followed by foot drop and later with wasting and weakness of the small hand muscles. Neurophysiological tests determine the degree of demyelination and axonal loss. No treatment is available and patients are advised to avoid local trauma to peripheral nerves at key points, for example the ulnar nerve at the elbow and the common peroneal nerve at the knee.

CASE REPORT

A commercial airline pilot reported to the UK Civil Aviation Authority a diagnosis of CMT disease at the age of 53 after his brother had recently been diagnosed with the same condition. Genetic testing showed that he had the dominantly inherited demyelinating X-linked CMTX1 variant due to mutations in GJB1 on chromosome Xq13.1, causing loss of the normal

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connexin 32 gene function.[2] Over 260 distinct mutations in this gene have been identified.[8] CMTX may be demyelinating, mixed, or axonal and dominant or recessive.[5] This form of CMT is about 7–12% of all cases. The first signs usually appear in childhood, but there may be no significant disability for several decades, as in this case.

There was a history of high arched feet in childhood, but apart from some difficulty with running and sports at school, the pilot had experienced no other problems. However, in his forties, he started to develop a very slowly progressive distal weakness in his feet and hands with pes cavus, foot drop, and wasting and weakness of the small hand muscles.

Clinical examination showed no abnormality in the cranial nerves. There was grade 2–4 weakness in the small hand muscles with normal power proximally. There was grade 2 weakness of ankle dorsiflexion; he could stand on both forefeet separately, but not on his heels. Pain sensation and joint position sense were very slightly impaired in the toes and two-point discrimination slightly widened on both index fingers. He had no sensory symptoms and was unaware of this minimal and subclinical sensory deficit. Electromyography and nerve conduction studies showed moderate slowing of nerve conduction velocities indicative of a predominately demyelinating sensorimotor neuropathy, with slight reduction in amplitude of the compound muscle action potentials showing some degree of axonal involvement.

DISCUSSION

The initial impression was that the extent of this pilot's neurological deficit would not be compatible with all the flying tasks of a commercial pilot, including those required in emergencies; but throughout his career there had been no history of any difficulties during flying and training and he had passed all the routine simulator flight checks to a high standard.

However, this does not necessarily ensure that his pattern of weakness would have no impact on flying, as a routine 6-mo simulator check does not test all tasks at one time, but a selection of tasks for each category of procedure, though all procedures are checked over a 3-yr cycle. Furthermore, fatigability is not tested in a routine simulator check and could be relevant in a pilot with weakness. As the condition is known to be progressive, a baseline measure would allow a regulator not only to determine suitability for medical recertification, but also to monitor progression over time. All pilots must meet the general medical standards for certification and the examination for this is different from a clinical examination in the neurology clinic.

A specifically tailored Medical Flight Test (MFT), choosing procedures from the full 3-yr cycle, was considered the most appropriate method to assess tasks which might be difficult for a pilot with this particular disability. All the tasks selected were taken from the well-developed and standardized simulator check protocols and there were no new or modified tasks designed for this assessment. The pilot had to pass all aspects of the MFT to an acceptable standard.

For the routine simulator checks required by EU Regulations and carried out by Type Rating Examiners, a Pass/Fail result is usually further graded against a number of competencies defined by each airline. These competency gradings could be used to monitor progression over time. While the nomenclature can vary from airline to airline, the following skills are usually described in standard operations manuals: technical skills (automatic flight control, manual flight control, and procedural knowledge); cognitive skills (situational awareness, decision making/problem solving, and workload management); and personal skills (communication, teamwork, leadership, and personal standards). Of these competencies, manual flight control, which includes all nonautomated aircraft controls on the ground and in the air, is of relevance in this pilot, for whom varying and precise levels of physical input are required. Automatic flight control is also important as significant switch and button selections and data inputs are required to ensure the Flight Management System and Autopilot Flight Director System are correctly programmed and managed. These inputs typically involve rotary switch and push button selections to control parameters such as speed, altitude, and autoflight modes on the Mode Control Panel and keyboard data entry on the three Control and Display Units.

Brief details of the MFT chosen for this pilot were as follows: B747-400 full flight simulator without a copilot, wet runway $<$ 3 mm. To test abnormally high control column forces, a 'Jammed Stabilizer' fault was applied during level flight at 320 kn. If this fault were to happen the aircraft would only be 'in trim' at the airspeed current at the time of the 'Jammed Stabilizer'. To prepare for a diversion and landing the pilot needs to reduce speed to configure the aircraft for landing by lowering the flaps and landing gear. As the stabilizer cannot be moved to trim the aircraft and thus relieve control column loads, the pilot must apply significant backward force on the control column while continuing to maintain accurate flight path control and prevent the aircraft from going into a dive. This force is substantially greater than any of the control column forces normally encountered and is tiring to maintain. The autopilot is degraded in this case and is not available for use.

High control column forces can also be encountered when the stabilizer moves uncommanded, putting the aircraft increasingly 'out of trim' (a 'Stabilizer Trim Runaway'). This requires the pilot to hold an increasing stick force for a sustained period of time until the movement is stopped by an electrical cut out switch and the aircraft put back 'in trim' with neutral stick forces.

To test the ability to operate overhead panel controls, an engine fire was simulated in the engine furthest away from the operating seat, requiring both extinguishing bottles. The fire switches are installed in the center of the overhead panel on the B747 and are 'T' handles which light up in the case of an engine fire. The initial action is to pull the switch out, which shuts down the engine and cuts the fuel supply, engine bleed air, hydraulic, and electrical power. The two available extinguishant bottles can then be discharged by turning the switch one way for 1 s against a light spring load and then the other way to

discharge a second bottle if required. The No. 4 engine fire switch is the furthest from the left-hand seat and therefore requires the longest reach.

To test the response to a Rejected Takeoff (RTO) at 100 kn, without the use of the Autobrake System, as a result of an engine failure, and with a maximum crosswind (25 kn) on a wet runway, the pilot has to apply the toe brakes evenly and simultaneously to maximum braking force with each foot when the rudder pedals are significantly displaced, one fore and one aft of neutral. The RTO maneuver was tested twice with an engine failure on each side separately to demonstrate the ability to control the aircraft with a power loss on each side.

To test the ability to control the aircraft after engine failure, a simulated failure of engine No. 1 (outer left) and No. 4 (outer right) between takeoff decision speeds V1 & V2 was performed separately. Above the V1 speed the takeoff must continue. The failure of an 'outboard engine' gives the most dramatic 'yaw' effect that must be corrected to keep the aircraft straight as a result of thrust asymmetry. Significant rudder input is required along with accurate pitch and roll control to assure the correct flight path both vertically and laterally.

Each of the foot operated toe brake pedals exerts a variable braking force to its 'own-side' main landing gear depending on the pressure applied. To test the ability to maneuver the aircraft while taxiing, the pilot must apply brake pressure to each brake pedal in turn to demonstrate the ability to exert asymmetric braking forces.

To test aircraft control with windshear on departure or approach, which can cause significant departure from the intended flight profile, requires prompt action and accurate manual flying skills, especially if the autopilot is not engaged. The B747 'Windshear Escape Maneuver' includes the following actions: disengage the autopilot, push either the takeoff or go around switches, which are an autopilot/autothrottle setting and which activates takeoff or go-around thrust, aggressively apply maximum thrust as this is quicker than the action of the takeoff/go around switches, disconnect the autothrottle, simultaneously roll the wings level and rotate toward an initial pitch attitude of +15°, and retract speed brakes.

The pilot successfully completed the Medical Flight Test with all the tests listed to a high standard. He only experienced a minor problem with the standby ignition switches located in the middle of the overhead panel that are easily accessible by both pilots. These rotary switches allow a standby power source to activate the engine igniters and may be required at initial start-up and are rarely, if ever, required in flight. The pilot was able to demonstrate that he could operate these switches, although occasionally requiring the use of both hands, particularly when tired. During normal flight operations he would have the assistance of the copilot. This pilot was, therefore, medically recertificated as fit for professional flying with a multicrew restriction.

Relying on a periodic medical examination or even a specialist examination may over- or underestimate a pilot's functional ability. The routine simulator flight check was modified by choosing items from the full 3-yr cycle to test the specific problems which may be faced by a pilot with this neurological deficit. For a fixed neurological deficit, a MFT may only be required once before a return to certification, but if the condition is likely to progress, as in this case, the MFT should be repeated at regular intervals to determine ongoing fitness for certification. The MFT should be performed at the time of the standard 6-mo simulator check, which was deemed to be reasonable in view of the very slow progression of this condition. The likelihood that this condition would cause a sudden incapacitation in flight is negligible. Neurophysiological examination (electromyography and nerve conduction studies) would provide objective and numerical evidence of disease progression, but there are no neurophysiological thresholds which would determine a pilot's ability to perform the tasks of professional flying. The procedure outlined here could be a model for the assessment of pilots with other neurological conditions.

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