Can J Anesth/J Can Anesth (2010) 57:248–255
DOI 10.1007/s12630-009-9244-1

CASE REPORTS/CASE SERIES

Anesthesia and myotonic dystrophy type 2: a case series
Anesthésie et dystrophie myotonique de type 2: une série de cas

Toby N. Weingarten, MD · Ryan E. Hofer, MMS · Margherita Milone, MD, PhD · Juraj Sprung, MD, PhD

Received: 31 July 2009 / Accepted: 2 December 2009 / Published online: 15 January 2010

Abstract

Background *Myotonic dystrophy type 2 (DM2) is a genetically distinct disorder that shares some phenotypical features of myotonic dystrophy type 1 (DM1). However, anesthetic management of patients with DM2 has not been described. The purpose of this study is to report the anesthetic management of a series of patients with DM2 and to describe their response to anesthesia.*

Methods *We performed a computerized search of the Mayo Clinic medical records database looking for patients with DM2 who underwent general anesthesia. The medical records were reviewed for anesthetic technique, medications used, and postoperative complications.*

Results *We identified 19 patients with DM2 who underwent 39 general anesthetics, 17 monitored anesthetic care cases, and two regional anesthetics. The patients exhibited normal responses to succinylcholine, nondepolarizing neuromuscular blockers, neostigmine, induction agents, and volatile anesthetics. Serious postoperative complications related to DM2 did not occur.*

Conclusion *In our series, patients with DM2 tolerated commonly used anesthetics without obvious complications, and they exhibited normal responses to muscle relaxants. These observations suggest that these medications may be used safely in patients with DM2.*

Résumé

Contexte *La dystrophie myotonique de type 2 (DM2) est un trouble génétiquement distinct qui partage certaines caractéristiques phénotypiques avec la dystrophie myotonique de type 1 (DM1). La prise en charge pour l'anesthésie de patients souffrant de DM2 n'a cependant pas été décrite. L'objectif de cette étude est de rendre compte de la prise en charge pour l'anesthésie d'une série de patients souffrant de DM2 et de décrire leur réponse à l'anesthésie.*

Méthode *Nous avons réalisé une recherche informatisée dans la base de données des dossiers médicaux de la Clinique Mayo pour identifier les patients souffrant de DM2 et ayant subi une anesthésie générale. Les dossiers médicaux de ces patients ont été révisés et l'on a noté la technique anesthésique et les médicaments utilisés ainsi que les complications postopératoires.*

Résultats *Nous avons identifié 19 patients souffrant de DM2 et ayant subi 39 anesthésies générales, 17 cas de sédation consciente, et deux cas d'anesthésie régionale. Des réponses normales à la succinylcholine, aux curares non dépolarisants, à la néostigmine, aux agents d'induction et aux agents anesthésiques volatils ont été observées chez ces patients. Il n'y a pas eu de complications postopératoires graves associées à la DM2.*

Conclusion *Dans notre série de cas, les patients souffrant de DM2 ont toléré les anesthésiques fréquemment utilisés sans complication évidente et ont montré des réponses normales aux curares. Ces observations suggèrent que ces médicaments peuvent être utilisés de façon sécuritaire chez des patients souffrant de DM2.*

T. N. Weingarten, MD (✉) · J. Sprung, MD, PhD
Department of Anesthesiology, Mayo Clinic College of Medicine, 200 First Street SW, Rochester, MN 55905, USA
e-mail: weingarten.toby@mayo.edu

R. E. Hofer, MMS
Mayo Clinic College of Medicine, Rochester, MN 55905, USA

M. Milone, MD, PhD
Department of Neurology, Mayo Clinic College of Medicine, Rochester, MN 55905, USA

Myotonic dystrophy type 2 (DM2) is a rare autosomal-dominant multisystem disorder characterized by proximal muscle weakness, myotonia, precocious cataracts, muscle pain, and muscle stiffness.[1,2] Though it shares some phenotypical features of classic or type 1 myotonic dystrophy (DM1), it is a genetically distinct disorder and recognized as a separate entity.[1–6] Anesthetic management of patients with DM2 has not been described, and it is unknown if these patients may have similar abnormal responses to anesthetic agents as do patients with DM1, for example, myotonic responses to succinylcholine,[7–10] increased sensitivity to nondepolarizing muscle relaxants,[3] myotonic responses to neostigmine,[3] or increased sensitivity to anesthetic agents.[11–14] To determine if patients with DM2 have similar atypical responses to anesthesia, we used the Mayo Clinic medical records database to identify patients with DM2 who underwent anesthesia, and we reviewed their anesthetic course.

Methods

After obtaining Mayo Clinic, Rochester, Minnesota, Institutional Review Board approval, a manual and computerized search of the Mayo Clinic Rochester medical records database from 2000 through 2008 was conducted to identify patients who were diagnosed with DM2 by Mayo clinic neurologists and who underwent anesthetic care at Mayo Clinic, Rochester, Minnesota**.** The medical records of these patients were then reviewed to determine if they had received anesthetic care. The anesthetic records were reviewed by one of the authors (R.E.H.); data were entered into a standardized data collection form, and all questionable entries were discussed with the senior author (T.N.W.). We reviewed demographics (age, gender), anesthetic techniques, responses to neuromuscular blocking agents and neostigmine, hemodynamic variability (blood pressure and/or heart rate episodes above or below 30% measured before anesthesia induction), hemodynamic instability (need for any pressor/chronotropic drugs), the occurrence of significant arrhythmias (new onset atrial fibrillation/flutter, tachycardias, complete heart block, etc.), and intraoperative body temperature. We also reviewed the records of the postoperative period for signs of delayed anesthetic recovery, myotonia, residual muscle weakness, need for prolonged ventilatory support or tracheal reintubation, and any note of postoperative respiratory or other complications. Only descriptive statistics were used.

Results

From January 1, 2000 to December 31, 2008, neurologists at the Mayo Clinic identified 74 patients with DM2, 61 by genetic confirmation. Nineteen (n = 19) patients with DM2 underwent anesthetic care (Table 1). Seventeen (n = 17) of these patients were symptomatic. One adult patient was asymptomatic but was found to have DM2 during an evaluation of elevated serum creatine kinase. One patient, a ten-yr-old female, was also asymptomatic but had genetically confirmed DM2. All cases were genetically confirmed except for one patient with symptomatic DM2 whose sister was genetically confirmed DM2. Six patients were diagnosed with DM2 after their surgical procedure.

Table 1 Demographic and other characteristics of patients with DM2

Characteristics	
Age at surgery (yr)*	56 (41, 60)
Gender (n)	
Female	14
Male	5
Duration of symptoms (yr)*	3 (1, 11)
Procedures (n)	
Otolaryngological and oral procedures	11
Orthopedic/plastics	10
General/urologic	8
Electroconvulsive therapy	18
Ophthalmologic	1
Radiologic	10
Symptoms/Findings (n)	
Muscle weakness	17
Clinical myotonia	11
Flaccid dysarthria	2
Cataracts	9
Associated co-morbid conditions (n)	
Diabetes mellitus	4
Heart conduction abnormalities	5
Cardiomyopathy	1
Prior aspiration pneumonia	1

* Median (25%, 75% quartiles); n = number of patients

All adult patients were assigned ASA physical status II or III. Seventeen of the adult patients had proximal muscle weakness, and in seven of these patients, the weakness was moderate. Eleven patients had mild clinical signs of myotonia, and in five of these patients, myotonia was moderate. Two of the patients with moderate myotonia and weakness also had flaccid dysarthria, while another patient with moderate myotonia had a history of aspiration pneumonia. Only one patient had decreased left ventricular function with an ejection fraction of 25%, but rather than DM2-associated cardiomyopathy, this decreased function was attributed primarily to the presence of significant coronary artery disease and prior chemotherapy to treat non-Hodgkin's lymphoma. Five patients (26.3%) had abnormal conduction patterns on their electrocardiograms: one

chronic atrial fibrillation, two right bundle branch blocks, one non-specific intraventricular conduction delay, and one left anterior fascicular block. Four patients (21%) in this series had diabetes mellitus. Cognitive function, as assessed clinically by a Mayo clinic neurologist, was fully preserved in 16 adult patients and they all lived independently. One patient, a 60-yr-old female with co-morbid Parkinson's disease, had severe psychiatric disorders requiring electroconvulsive therapy and nursing home care. Cataracts were common (n = 9, 47.4%).

The adult patients underwent 49 procedures that required anesthesia: 37 general anesthetics, ten cases that required monitored anesthesia care, one spinal anesthetic, and one infraclavicular nerve block (Table 2). Aside from using etomidate to induce general anesthesia in the patient with the reduced left ventricular ejection fraction, propofol or sodium thiopental were used to induce general anesthesia in all other cases. Succinylcholine was used to facilitate tracheal intubation for seven anesthetics in seven patients, and increased skeletal muscle tone or unusual effects on muscle relaxation were not noted. Nondepolarizing muscle relaxants were used in 32 cases and reversed with neostigmine in 24 cases with normal responses. Muscle relaxant administration was titrated according to train-of-four muscle contractions in response to peripheral neuromuscular stimulation**.** Review of medical records revealed no indication of atypical or prolonged response to muscle relaxants or abnormal responses to neostigmine. No cases of significant hypotension or perioperative cardiac dysrhythmias were noted. Intraoperative body temperature was easily controlled with the use of forced-air warming blankets. Perioperative glucose levels were not consistently obtained for patients in this series. In the non-diabetic adults, these values were obtained in five cases, with the highest value being 140 $mg{\cdot}dL^{-1}$. The highest plasma glucose level among the adult type 2 diabetics was 247 $mg{\cdot}dL^{-1}$. No adult patients were administered insulin. All patients had unremarkable anesthetic recovery without significant postoperative cardiovascular or pulmonary complications. Five patients who received general anesthesia had more than a two-hour recovery room time for the following reasons: two patients had poorly controlled pain and nausea; two patients had to wait for a room assignment; and one patient was somnolent following a 658 min general anesthetic for a plastic surgical procedure. However, the tracheas of these patients were extubated in the operating room at the end of the surgical procedures. Another patient who received a spinal anesthetic remained in the recovery room for 152 min until the nerve block receded.

The ten-yr-old female presented for surgical management of a ruptured appendix that required general anesthesia for two exploratory laparotomies as well as seven radiologic procedures, i.e., sonogram, requiring monitored anesthesia care. The patient had normal responses to anesthesia care, including the administration of propofol for induction, rocuronium and atracurium for muscle relaxation, and reversal with neostigmine. She developed postoperative hyperglycemia (484 $mg{\cdot}dL^{-1}$) and was subsequently diagnosed as having diabetes mellitus type 1. Her plasma insulin level was low normal (4.4 $UIU{\cdot}mL^{-1}$; range 1.4–14 $UIU{\cdot}mL^{-1}$), which suggested that her diabetes was due to low insulin production rather than insulin resistance. From her evaluation for hyperglycemia, it was learned that she had a family history of "mild" myotonic dystrophy. She underwent genetic testing and was found to have DM2. However, save for her diabetes, she was asymptomatic for DM2.

Discussion

Myotonic dystrophy type 2 (DM2) is an autosomal-dominant multisystem disorder characterized by proximal muscle weakness, myotonia, myalgias, muscle stiffness, and precocious cataracts, and it may also affect the cardiovascular and endocrine systems.[1,2] Until the early 1990s, myotonic dystrophy was considered to be a homogeneous disease, but in the mid 1990 s, a subset of patients were identified who differed genetically[15] and phenotypically from patients with DM1 (Table 3).[1] Important distinctions of DM2 patients are the predominance of proximal muscle weakness and a milder disease course compared with DM1 patients.[1,2]

Our DM2 patients tolerated commonly used anesthetic medications satisfactorily and exhibited normal response to a variety of muscle relaxants, including succinylcholine and several nondepolarizing relaxants. Also, reversing the effects of muscle relaxants with neostigmine was uneventful. With the exception of the ten-yr-old female developing hyperglycemia, our patients' intraoperative and postoperative periods were unremarkable. The cases where recovery room stay was prolonged were the result of factors other than excessive sensitivity to anesthesia. From our limited series, it appears that these medications are tolerated well in patients with DM2 and, thus, may be used safely. This is in contrast to patients with classic DM1 where various adverse effects due to muscular and extramuscular involvement were reported, including an exaggerated response to sodium pentothal[11,14] and propofol,[12,13] a myotonic reaction to succinylcholine,[7–10] unpredictable and inadequate muscle relaxant reversal, increased muscle weakness and myotonia to neostigmine,[3] and prolonged apnea following sodium pentothal.[14]

Diabetes mellitus is common in DM2 and is a result of altered insulin receptors on skeletal muscle that contribute to insulin resistance in this disorder.[16] In our series, diabetes mellitus was present in four adult patients. There

Table 2 Surgical and anesthetic characteristics of patients with myotonic dystrophy type 2 undergoing anesthesia

Patient	Age @ surgery	Age @ diagnosis	Type of surgery	Anesthetic technique	Anesthetic duration (min)	Induction agent, dose (GA only)	SUX use, dose (mg)	NDMR type, dose (mg)	Neostigmine use, dose (mg)	Maintenance agent, dose (% or mcg·kg^{-1}·min^{-1})	N_2O (%)	PACU stay duration (min)
1	62	67	Gen	GA	129	P, 150	120	Cis, 10	5	Iso, 1.2	70	108
2	76	74	Ortho	GA	207	P, 180				Sev, 1.4	60	B/P
3	1. 49	45	1. ENT	1. GA	1. 149	1. P, 200		1. Vec, 8	1. Not used	1. Sev, 2.3		1. 45
	2. 51		2. Cath	2. MAC	2. 125					2. Mid/Fent		2. 34
4	56	61	Dent	GA	145	P, 150	120			Sev, 1.2	60	136
5	1. 41	41	1. Dent	1. MAC	1. 41			1.		1. Mid/Fent		1. B/P
	2. 41		2. Dent	2. GA	2. 92	2. P, 150		2. Vec, 5	2. 4	2. Des, 6.0		2. 62
	3. 41		3. ENT	3. MAC	3. 46			3.		3. Mid/Fent		3. B/P
	4. 43		4. ENT	4. GA	4. 161	4. P, 180		4. Roc, 7	4. Not used	4. P, 120	4. 70	4. 27
	5. 43		5. Gen	5. GA	5. 108	5. P, 200		5. Cis, 12	5. 4	5. P, 100, Sev, 0.8	5. 50	5. 33
6	34	39	Dent	GA	146	P, 200	150	Vec, 1	Not used	Iso, 0.8	66	36
7	1. 73	71	1. ENT	1. GA	1. 180	1. P, 200		1. Miv, 6	1. Not used	1. Iso, 1.0	1. 30	1. 75
	2. 77		2. Gen	2. GA	2. 172	2. P, 70		2. Vec, 5	2. 3	2. Sev, 1.3	2. 60	2. 173
	3. 77		3. Ortho	3. GA	3. 118	3. P, 120	3. 120			3. Sev, 1.2	3. 55	3. 80
8	1–18: 60	60	1–18: ECT	1–18: GA	1–18: <30	STP, 175–250		Ata, 20–30	4, all cases	None		1–18: <30
9	40	41	Ortho	Regional	148					None		B/P
10	51	51	ENT	GA	304	P, 80		Miv, 10	Not used	Des, 6.1		167
11	1. 52	50	1. Gen	1. GA	1. 225	1. P, 200	1. 140	1. Vec, 8	1. 5	1. Iso, 1.2		1. 90
	2. 54		2. Pls	2. GA	2. 658	2. P, 140		2. Vec, 14	2. Not used	2. Iso, 0.7	2. 60	2. 128
12	77	77	Gen	MAC	59					Mid/Fent		44
13	73	72	Ortho	Spinal	157					P, 30		152
14	1. 38	36	1. Gen	1. GA	1. 55	1. P, 200		1. Vec, 5	1. Not used	1. P, 25	1. 70	1. 39
	2. 40		2. Dent	2. MAC	2. 90					2. Mid/Fent		2. 9
	3. 41		3. Cath	3. MAC	3. 49					3. Mid/Fent		3. 47
	4. 42		4. Gen	4. GA	4. 73	4. P, 200				4. Iso, 1.1	4. 50	4. 73
	5. 43		5. Dent	5. MAC	5. 86					5. P/Mid/Fent		5. 19
	6. 44		6. Cath	6. MAC	6. 116					6. Mid/Fent		6. 46
15	1. 65	66	1. Gen	1. MAC	1. 32					1. P, 10		1. B/P
	2. 66		2. Ortho	2. GA	2. 163	2. STP, 300		2. Cis, 26	2. Not used	2. Iso, 0.5	2. 50	2. 63
16	54	54	Uro	GA	164	P, 150	160	Cis, 16	Not used	Iso, 0.7		77
17	63	64	Ophth	MAC	111					P, 40 mg bolus		28
18	71	70	Ortho	GA	152	E, 20	160			Sev, 2.0	35	174

Table 2 continued

Patient	Age @ surgery	Age @ diagnosis	Type of surgery	Anesthetic technique	Anesthetic duration (min)	Induction agent, dose (GA only)	SUX use, dose (mg)	NDMR type, dose (mg)	Neostigmine use, dose (mg)	Maintenance agent, dose (% or mcg·kg^{-1}·min^{-1})	N_2O (%)	PACU stay duration (min)
19	1–9: 10	12	1. Gen	1. GA	1. 195	1. STP, 150		1. Roc, 25	1. 0.5	1. Sev, 2.9		1. B/P
			2. Gen	2. GA	2. 152	2. P, 90		2. Ata, 2	2. Not used	2. Iso, 1.5		2. 57
			3. IR	3. MAC	3. 185					3. P/Mid/Fent		3. 38
			4. Gen	4. MAC	4. 213					4. P, 30		4. B/P
			5. IR	5. GA	5. 122	5. P, 50				5. Sev, 2.2		5. B/P
			6. IR	6. MAC	6. 113					6. P/Mid/Fent		6. 39
			7. IR	7. MAC	7. 176					7. P, 10		7. 57
			8. IR	8. MAC	8. 167					8. P/Fent		8. 47
			9. IR	9. GA	9. 104	9. SEV				9. Sev, 2.6		9. B/P

Gen = general surgical procedure; Ortho = orthopedic surgical procedure; ENT = otolaryngologic surgical procedure; Cath = cardiac catheterization; Dent = dental surgical procedure; ECT = electroconvulsive therapy; Pls = plastic surgical procedure; Uro = urological surgical procedure; IR = interventional radiology procedure; GA = general anesthesia; MAC = monitored anesthetic care; P = propofol; STP = sodium thiopental; SUX = succinylcholine; NDMR = nondepolarizing muscle relaxants; Cis = cisatracurium; Vec = vecuronium; Roc = rocuronium; Miv = mivacurium; Ata = atracurium; Iso = isoflurane; Sev = sevoflurane; Des = desflurane; Mid = midazolam; Fent = fentanyl; PACU = postanesthesia care unit (recovery); B/P = bypass recovery room

were no clinically significant elevated plasma glucose levels among non-diabetic adult patients in this series; however, perioperative glucose levels were not obtained consistently, and hyperglycemia may have been unrecognized in these cases. The manifestation of diabetes in the ten-yr-old female in the postoperative period may have been coincidental, as she had low levels of plasma insulin, which suggests her diabetes was not secondary to insulin resistance. Regardless, because of the propensity of these patients to have insulin resistance, anesthesiologists should monitor glucose levels closely during the perioperative period.

Cardiac involvement appears to be more benign in DM2 patients compared with DM1 patients, although a case of severe cardiomyopathy has been reported.[17] The most frequent cardiac symptoms in DM2 patients are palpitation, intermittent tachycardia, and syncopal spells.[2] Conduction abnormalities and intraventricular and atrioventricular blocks were also described,[2] and were present in five of our patients. It is well established that sudden death can occur as a consequence of cardiac-conduction abnormalities in the DM1 patients.[18] However, it is not known whether the same risk applies to DM2 patients. In one series of 297 DM2 patients, sudden death occurred in four patients. Three of the patients had no prior cardiac symptomatology, and one patient had a history of heart failure.[19] Cardiac histopathology showed dilated cardiomyopathy in all four patients and conduction system fibrosis in two patients with pathogenetic DM2-specific ribonuclear inclusions demonstrable in cardiomyocytes.[19] In DM2 patients without overt cardiac disease, magnetic resonance spectroscopy of the left ventricular myocardium has found evidence of the existence of subclinical cardiomyopathy.[20] In our series, only one patient had clinically significant cardiomyopathy, which was attributed to chemotherapy and coronary artery disease. In retrospect, however, DM2-associated cardiomyopathy cannot be excluded. While cardiac rhythm disturbances have been described in patients with DM2, these patients do not have major abnormalities of cardiovascular autonomic function.[21] We still have limited knowledge in assessing the cardiac risk of DM2 patients undergoing anesthesia, and it is unclear whether their risk of a perioperative cardiovascular event differs from the general population.

Gastrointestinal symptoms are highly prevalent in DM2 patients.[22] Dysphagia for liquids and solid food, abdominal pain, and constipation were significantly more common among DM2 patients than among healthy controls and were comparable with DM1 patients. When present, dysphagia in DM2 patients is generally mild but becomes more severe with age.[22] Our 40-yr-old patient who had a history of aspiration pneumonia had more severe symptoms compared with other patients in our series. Since advanced myotonic

Table 3 Comparison of characteristics between DM1 and DM2

	DM 1	DM 2
Genetic Characteristics[15]		
Inheritance	Autosomal dominant	Autosomal dominant
Gene	DMPK (Protein kinase)	ZNF9 (Zinc finger protein 9)
Locus	19q13	3q21
Mutation	CTG-repeat expansion	CCTG-repeat expansion
Clinical Characteristics		
Onset	birth, childhood, or adult onset	Adult onset[1,2]
Cognitive Function	Congenital form: mental retardation[25] Adult onset: Age related decline of frontal and temporal cognitive function[25–27]	Age related decline of executive cognitive function[26,27]
Facial Abnormalities		
Cataracts	Posterior subcapsular iridescent cataracts common	Posterior subcapsular iridescent cataracts common[2]
Frontal Balding	Common	Less common[1]
Endocrine Dysfunction		
Diabetes (Insulin Resistance)	Common	Common[2]
Male Hypogonadism	Common	Less common[1,28]
Cardiac Dysfunction		
Conduction Abnormalities	Bradyarrhythmias[29] Tachyarrhythmias[18]	May be present[2,30,31]
Cardiomyopathy	Dilated Cardiomyopathy	Dilated Cardiomyopathy[2,17]
Death	Sudden cardiac death is well recognized[18,29]	Sudden cardiac death has been described[2,19]
Gastrointestinal Dysfunction		
Dysphagia[32]	Severe	Mild
Constipation[22]	Common	Common
Skeletal Muscle Features[1,2,33,34]		
Muscle Weakness	Mild to severe Distal > Proximal	Mild to moderate Proximal > Distal
Clinical myotonia	Prominent	Less prominent or absent
Myalgia	Uncommon	Prominent
Elevated creatine kinase	Common	Common
Responses to Anesthetic Medications		
Sodium Pentothal	Increased sensitivity[11,14]	Normal response*
Propofol	Increased sensitivity[12,13]	Normal response*
Succinylcholine	Myotonic Reactions[7–10]	Normal response*
Neostigmine	Inadequate reversal and myotonic reactions[3]	Normal response*

* Normal responses to these agents were observed in this case series. DM1 = myotonic dystrophy type 1; DM2 = myotonic dystrophy type 2

conditions may increase the risk for aspiration, preemptive measures need to be implemented perioperatively.

Clinical presentation of DM2 with body stiffness and muscle pains may resemble fibromyalgia. One study found that 3.2% of patients with fibromyalgia were tested positive for DM2 mutation.[23] Physicians, especially anesthesiologists evaluating these patients in a pain clinic, should be aware of possible overlap in the clinical presentation of these two distinct disorders and should focus their attention on subtle signs and symptoms that may indicate the presence of DM2.

In one series, 21% of women had the initial symptoms of DM2 during pregnancy and worsening of symptoms in subsequent pregnancies.[24] Thirteen percent of 96 pregnancies ended as early miscarriages, and 4% ended as late miscarriages. Preterm labour occurred in 50% of the pregnancies resulting in 27% preterm deliveries in women with overt DM2.[24] Presently, there is no evidence that regional anesthesia or labour epidural analgesia can affect outcomes of pregnancy in patients with DM2.

Since DM2 is a very rare disorder, it is impossible to conduct large-scale studies to assess the safety of

anesthesia in these patients. Thus, we must rely on experience gained from reports of individual cases or case series. Our case series has all the inherent limitations of a retrospective observational study, with the possibility that subtle complications or abnormal responses to anesthetic medications may not have been captured by our electronic medical records. It is also unclear whether our favourable results were secondary to a modification of typical anesthetic management by the attending anesthesiologist to account for the fact that the patients had DM2. In the instance of the patient who underwent multiple electroconvulsive treatments, there was a clear deviation from standard anesthetic care at our institution. In that case, the treating anesthesiologists used atracurium as the muscle relaxant rather than succinylcholine, which is the norm. Though speculative, the decision to use a nondepolarizing muscle relaxant was probably secondary to concerns about the use of succinylcholine in a patient with myotonic dystrophy. However, the diagnosis of DM2 in six patients was made following the surgery, so it can be assumed that these patients received standard care. Therefore, our results should be interpreted with a degree of caution, and further observational studies of patients with DM2 are necessary to make definitive conclusions regarding the safety of anesthesia.

In conclusion, in our series of 19 patients and 58 anesthetics, we could not detect anesthetic complications that could be attributed to DM2. Thus, it appears that commonly used anesthetic induction agents, i.e., succinylcholine, nondepolarizing muscle relaxants, and neostigmine are well tolerated in patients with DM2 and may be used safely. Although the clinical course of DM2 is more benign than that of DM1, anesthesiologists should be aware that patients with DM2 have a propensity to develop diabetes mellitus, dysphagia, cardiac conduction abnormalities, and cardiomyopathies.

Acknowledgements The authors would like to thank Drs. Jasper Daube and Nathan Young for their assistance in identifying patients with DM2.

Financial support This work was entirely supported by the Department of Anesthesiology, Mayo Clinic, Rochester, MN. The authors have no commercial or non-commercial affiliations or other associations that present or could be construed to present a conflict of interest.

References

1. *Finsterer J*. Myotonic dystrophy type 2. Eur J Neurol 2002; 9: 441–7.
2. *Day JW*, *Ricker K*, *Jacobsen JF*, *et al*. Myotonic dystrophy type 2: molecular, diagnostic and clinical spectrum. Neurology 2003; 60: 657–64.
3. *Buzello W*, *Krieg N*, *Schlickewei A*. Hazards of neostigmine in patients with neuromuscular disorders. Report of two cases. Br J Anaesth 1982; 54: 529–34.
4. *Ranum LP*, *Rasmussen PF*, *Benzow KA*, *Koob MD*, *Day JW*. Genetic mapping of a second myotonic dystrophy locus. Nat Genet 1998; 19: 196–8.
5. *Ricker K*. Myotonic dystrophy and proximal myotonic myophathy. J Neurol 1999; 246: 334–8.
6. *Udd B*, *Krahe R*, *Wallgren-Pettersson C*, *Falck B*, *Kalimo H*. Proximal myotonic dystrophy–a family with autosomal dominant muscular dystrophy, cataracts, hearing loss and hypogonadism: heterogeneity of proximal myotonic syndromes? Neuromuscul Disord 1997; 7: 217–28.
7. *Thiel RE*. The myotonic response to suxamethonium. Br J Anaesth 1967; 39: 815–21.
8. *Orndahl G*. Myotonic human musculature: stimulation with depolarizing agents. I. Mechanical registration of the effects of acetylcholine and choline. Acta Med Scand 1962; 172: 739–51.
9. *Orndahl G*. Myotonic human musculature: stimulation with depolarizing agents. II. A clinico-pharmacological study. Acta Med Scand 1962; 172: 753–65.
10. *Orndahl G*, *Stenberg K*. Myotonic human musculature: stimulation with depolarizing agents. Mechanical registration of the effects of succinyldicholine, succinylmonocholine and decamethonium. Acta Med Scand Suppl 1962; 389: 3–29.
11. *Dundee JW*. Thiopentone in dystrophia myotonia. Curr Res Anesth Analg 1952; 31: 257–62.
12. *Speedy H*. Exaggerated physiological responses to propofol in myotonic dystrophy. Br J Anaesth 1990; 64: 110–2.
13. *White DA*, *Smyth DG*. Exaggerated physiological responses to propofol in myotonic dystrophy. Br J Anaesth 1990; 64: 758–9.
14. *Aldridge LM*. Anaesthetic problems in myotonic dystrophy. A case report and review of the Aberdeen experience comprising 48 general anaesthetics in a further 16 patients. Br J Anaesth 1985; 57: 1119–30.
15. *Liquori CL*, *Ricker K*, *Moseley ML*, *et al*. Myotonic dystrophy type 2 caused by a CCTG expansion in intron 1 of ZNF9. Science 2001; 293: 864–7.
16. *Savkur RS*, *Philips AV*, *Cooper TA*, *et al*. Insulin receptor splicing alteration in myotonic dystrophy type 2. Am J Hum Genet 2004; 74: 1309–13.
17. *Wahbi K*, *Meune C*, *Bassez G*, *et al*. Left ventricular noncompaction in a patient with myotonic dystrophy type 2. Neuromuscul Disord 2008; 18: 331–3.
18. *Groh WJ*, *Groh MR*, *Saha C*, *et al*. Electrocardiographic abnormalities and sudden death in myotonic dystrophy type 1. N Engl J Med 2008; 358: 2688–97.
19. *Schoser BG*, *Ricker K*, *Schneider-Gold C*, *et al*. Sudden cardiac death in myotonic dystrophy type 2. Neurology 2004; 63: 2402–4.
20. *Schneider-Gold C*, *Beer M*, *Kostler H*, *et al*. Cardiac and skeletal muscle involvement in myotonic dystrophy type 2 (DM2): A quantitative 31P-MRS and MRI study. Muscle Nerve 2004; 30: 636–44.
21. *Flachenecker P*, *Schneider C*, *Cursiefen S*, *Ricker K*, *Toyka KV*, *Reiners K*. Assessment of cardiovascular autonomic function in myotonic dystrophy type 2 (DM2/PROMM). Neuromuscul Disord 2003; 13: 289–93.
22. *Tieleman AA*, *van Vliet J*, *Jansen JB*, *van der Kooi AJ*, *Borm GF*, *van Engelen BGM*. Gastrointestinal involvement is frequent in myotonic dystrophy type 2. Neuromuscul Disord 2008; 18: 646–9.
23. *Auvinen S*, *Suominen T*, *Hannonen P*, *Bachinski LL*, *Krahe R*, *Udd B*. Myotonic dystrophy type 2 found in two of sixty-three persons diagnosed as having fibromyalgia. Arthritis Rheum 2008; 58: 3627–31.

24. *Rudnik-Schoneborn S*, *Schneider-Gold C*, *Raabe U*, *Kress W*, *Zerres K*, *Schoser BG*. Outcome and effect of pregnancy in myotonic dystrophy type 2. Neurology 2006; 66: 579–80.
25. *Modoni A*, *Silvestri G*, *Pomponi MG*, *Mangiola F*, *Tonali PA*, *Marra C*. Characterization of the pattern of cognitive impairment in myotonic dystrophy type 1. Arch Neurol 2004; 61: 1943–7.
26. *Gaul C*, *Schmidt T*, *Windisch G*, *et al*. Subtle cognitive dysfunction in adult onset myotonic dystrophy type 1 (DM1) and type 2 (DM2). Neurology 2006; 67: 350–2.
27. *Sansone V*, *Gandossini S*, *Cotelli M*, *Calabria M*, *Zanetti O*, *Meola G*. Cognitive impairment in adult myotonic dystrophies: a longitudinal study. Neurol Sci 2007; 28: 9–15.
28. *Moxley RT 3rd, Meola G, Udd B, Ricker K*. Report of the 84th ENMC workshop: PROMM (proximal myotonic myopathy) and other myotonic dystrophy-like syndromes: 2nd workshop. 13-15th October, 2000, Loosdrecht, The Netherlands. Neuromuscul Disord 2002; 12: 306–17.
29. *Mathieu J*, *Allard P*, *Potvin L*, *Prevost C*, *Begin P*. A 10-year study of mortality in a cohort of patients with myotonic dystrophy. Neurology 1999; 52: 1658–62.
30. *Meola G*, *Sansone V*, *Marinou K*, *et al*. Proximal myotonic myopathy: a syndrome with a favourable prognosis? J Neurol Sci 2002; 193: 89–96.
31. *von zur Muhlen F*, *Klass C*, *Kreuzer H*, *Mall G*, *Giese A*, *Reimers CD*. Cardiac involvement in proximal myotonic myopathy. Heart 1998; 79: 619–21.
32. *Tieleman AA*, *Knuijt S*, *van Vliet J*, *de Swart BJ*, *Ensink R*, *van Engelen BG*. Dysphagia is present but mild in myotonic dystrophy type 2. Neuromuscul Disord 2009; 19: 196–8.
33. *Milone M*, *Batish SD*, *Daube JR*. Myotonic dystrophy type 2 with focal asymmetric muscle weakness and no electrical myotonia. Muscle Nerve 2009; 39: 383–5.
34. *Young N, Milone M, Daube JR*. Absent or unrecognized electromyographic myotonia in myotonic dystrophy type 2. Neurology 2009; 72 Suppl 3: A823.