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Comparison between clinical and autopsy diagnoses in a cardiology hospital

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Background: A few recent studies have evaluated diagnostic accuracy by comparison between clinical and autopsy diagnoses in a hospital specialising in cardiology.

Methods: 406 consecutive autopsy cases during 2 years were studied. Patients were aged 47.4 ± 28.4 years; 236 (58.1%) were men and 170 (41.9%) women. Diagnostic comparison was categorised in classes I to V (I, II, III and IV: discrepancy in decreasing order of importance regarding therapy and prognosis; V: concordance). Categorisation was ranked on the basis of the highest degree of discrepancy. Statistical analysis was performed with the χ^2 test and stepwise logistic regression.

Results: Each age increase of 10 years added 16.2% to the risk of the diagnostic comparison to be categorised in classes I and II (major discrepancy) in comparison to classes III, IV and V (OR 1.16, 95% CI 1.07 to 1.27, $p < 0.001$). By contrast, admission to intensive care units decreased the risk of categorisation in classes I and II by 47% (OR 0.53, 95% CI 0.32 to 0.85, $p = 0.009$). The most frequent diagnostic discrepancy occurred for pulmonary embolism: 30 out of 88 (34.1%) diagnoses in classes I and II. The concordance rate was 71.1% for acute myocardial infarction, 75% for aorta dissection, 73.1% for infective endocarditis and 35.2% for pulmonary embolism.

Conclusion: Age and hospital ward influenced the distribution of diagnostic discrepancy or concordance between clinical and autopsy diagnoses. The lower discrepancy rate for myocardial infarction and infective endocarditis may be related to the fact that the study was carried out in a specialist hospital.

Diagnostic accuracy may be evaluated by the comparison between clinical and autopsy diagnoses. In studies that compared these results carried out two decades apart, the discrepancies between clinical and autopsy diagnoses persisted in spite of progress in medical skills and technology.^{1–2} Further, the diagnoses of cardiovascular diseases were the most frequently missed diagnoses in the patients that died in a general hospital experience,³ including pulmonary embolism, myocardial infarction and infective endocarditis.⁴

A few recent studies have evaluated the comparison of clinical and autopsy diagnoses in a university hospital specialising in cardiology with a large outpatient clinic and emergency department as well as an active department of pathology. In spite of the ongoing progress in medical knowledge and technology, diagnostic challenges occur in the care of the patients.^{1–2,5}

The objective of this study was to compare clinical and autopsy diagnoses in classes of diagnostic comparison in a hospital devoted mainly to the care of patients with heart disease in a consecutive autopsy series over two years.

METHODS

Study setting

The study was performed in a 428-bed public university tertiary care hospital devoted mainly to cardiology, that also provides primary and secondary care, and includes a large outpatient practice and an emergency department. Death rates were 960 (8.2%) out of 11 715 admissions in 2001 and 926 (7.4%) out of 12 568 admissions in 2002.

Autopsies

Autopsies were performed by seven pathologists, all but one devoted to cardiovascular diseases, after medical request and

family consent following ethical and legal requirements. The brain, heart, lungs, kidneys, liver and spleen were examined in all cases. Complete autopsy (removal of the thoracic and abdominal organs en bloc) was performed if there was evidence of involvement of other organs, and in cases of congenital heart disease. Sampling for histology guided by the clinical and macroscopical findings was performed at least for the heart, lungs, kidneys and liver.

The autopsy rate was 23.6% (227 autopsies out of 960 deaths) in 2001 and 22.4% (207 out of 926 deaths) in 2002. The autopsy rate of patients who died in the emergency department was 163 of 383 (42.6%) deaths, in the intensive care units was 176 of 1151 (15.3%) deaths, and in other hospital wards was 95 of 352 (27%) deaths. Complete autopsy was performed in 150 (34.6%) of 434 cases.

Comparison of autopsied and non-autopsied cases in the hospital (Sao Paulo City Authority Program for the Study of Mortality database) revealed that the autopsy rate was higher in children under 1 year of age (14.9% vs 5.1%;) and lower in patients older than 71 years (27.6% vs 44%) ($\chi^2 = 93.1$; $p < 0.001$) and did not differ significantly relative to sex. For cardiovascular diseases, the autopsy rate was higher in patients with valvular heart diseases (26.6% vs 16.2%) and cardiomyopathies (14.8% vs 8%) and lower in patients with ischaemic heart disease (40.6 vs 53%) ($\chi^2 = 44.1$; $p < 0.001$). For non-cardiovascular diseases, the autopsy rate was higher in patients with congenital malformations (49.1% vs 23.2%) and lower in patients with diseases of the respiratory system (7.8% vs 23.9%) ($\chi^2 = 48.3$; $p < 0.001$).

Inclusion and exclusion criteria

All autopsy cases from 2001 and 2002 were studied. Twenty-eight cases (6.5% of the total 434 autopsies) were excluded due to limitations for retrieving and assessing the studied variables.

Study sample

A total of 406 patients were studied, of which 236 (58.1%) were male and 170 (41.9%) were female. The age of the patients ranged between 2 days and 92 years (mean 47.4; SD = 28.4). The age distribution was: <1 year in 60 (14.8%) patients, 1–10 years in 21 (5.2%) patients, 11–20 years in 15 (3.7%) patients, 21–30 years in 12 (3%) patients, 31–40 years in 26 (6.4%) patients, 41–50 years in 37 (9.1%) patients, 51–60 years in 53 (13%) patients, 61–70 years in 68 (16.7%) patients, 71–80 years in 84 (20.7%) patients, and over 80 years in 30 (7.4%) patients.

The duration of hospitalisation ranged from hours to 195 days (mean = 16 days; SD = 24.6 days).

One hundred and fifty-three patients aged 60.9 ± 19.7 years were hospitalised for 3.4 ± 7.8 days and died in the emergency department, 161 patients aged 38.5 ± 30.3 years were hospitalised for 25.8 ± 31.5 days and died in the intensive care units, and 92 patients aged 41.9 ± 30.3 years were hospitalised for 19.7 ± 19.7 days and died in other hospital wards.

Diagnostic criteria

- Clinical diagnoses for the purpose of this investigation followed previous studies^{4–6} and were defined as: (a) diagnoses retrieved from hospital records; (b) diagnoses informed in the autopsy request; (c) when treatment administered was specific to the point that a diagnosis might be devised in spite of not being literally written (anticoagulation for pulmonary embolism, recommended treatment regimen for tuberculosis).
- Autopsy diagnoses were made on the basis of macroscopic and microscopic examination.
- Diagnostic coding was made following the International Classification of Diseases, tenth revision.⁷
- The diagnosis of each case was reviewed by at least three of four investigators (RS, AJM, PSG, ATY) according to the adopted criteria. In the event of a disagreement, data were reviewed until a consensus was reached.

Comparison between clinical and autopsy diagnoses

The comparison was performed following a previously published categorisation¹ later modified.⁴

Diagnoses of diseases that were considered the cause of death were classified as major diagnoses and the diagnoses of other diseases were classified as minor diagnoses. The autopsy diagnosis of a disease in the absence of a previous clinical diagnosis was considered a discrepancy (whether a major discrepancy for the major diagnoses or a minor discrepancy for minor diagnoses).

The major discrepancies that were not clinically identified were categorised in classes I and II. Class I is a discrepant major diagnosis with potential adverse impact on survival and class II is a discrepant major diagnosis with equivocal impact on survival.

The minor discrepancies that were not clinically identified were categorised in classes III and IV. Class III is a discrepant minor diagnosable disease that was not directly related to the cause of death but was either symptomatic and should have been treated or would have eventually affected prognosis. Class IV is a discrepant non-diagnosable (occult) minor disease of possible genetic or epidemiological importance.

The concordant diagnoses were categorised in class V.

In the event of more than one diagnosis, categorisation might be performed in different classes in the same patient. In these cases, the patient was ranked in the class according to the discrepant diagnosis of a greater importance related to the potential influence on therapy and prognosis (class I > class II

> class III > class IV > class V). For instance, if a patient had two diagnoses of class V, one diagnosis of class IV and one diagnosis of class II, the comparison between clinical and autopsy diagnosis would categorise this patient as class II.

Studied variables

The comparison between clinical and autopsy diagnoses categorised in the classes above was evaluated relative to: (a) age; (b) sex; (c) duration of hospitalisation; (d) hospital ward where death occurred; (e) surgery (previous or during the admission in which death occurred). The comparison was performed according to the International Classification of Diseases.⁷

Statistical analysis

The comparison of proportions between groups was performed with the χ^2 test. Multivariate analysis was performed with stepwise logistic regression.^{8–9}

We compared patients with major discrepancies (classes I and II) relative to minor discrepancies (classes III and IV) and to concordance (class V). The Hosmer–Lemeshow test¹⁰ demonstrated that the model was well adjusted after stepwise variable selection.¹¹ P values <0.05 were considered significant.

Ethical aspects

The project was approved by the ethics review board of the hospital.

RESULTS

A total of 2884 autopsy diagnoses (mean 7.1 diagnoses per patient, SD = 2.5) were made in 406 cases. Most diagnoses (66.1%) were categorised in class V (table 1). The frequency of patients was higher (31.8%) in class IV (table 2).

The most frequent diagnoses were related to the circulatory system: 1645 diagnoses (table 1). Twenty-eight diagnoses (1.7%) were categorised in class I, 52 (3.2%) in class II, 86 (5.2%) in class III, 372 (22.6%) in class IV, and 1107 (67.3%) in class V. The diagnoses of diseases of circulatory system with frequency higher than 15 in this series with at least one diagnosis in class I are shown in table 3.

The classes of diagnostic comparison relative to the hospital ward where the patient died are shown in table 2.

Increasing age was associated with a higher frequency of patients with diagnoses categorised in classes I and II (fig 1; table 4). There was no significant difference relative to sex (table 4).

There was a significant difference in classes of diagnostic comparison relative to the hospital ward where the patient died (table 4); discrepancy rate (classes I and II) was less frequent in intensive care units (table 2). By contrast, the duration of hospitalisation and surgical treatment (either previous or in the last hospital admission before death) were not associated with a significant difference in the classes of diagnostic comparison.

The concordance rate in diseases of the circulatory system was 67.3%. The concordance rate was higher in patients with congenital heart diseases (table 1); 68 of 81 (84%) patients under 11 years of age had congenital heart diseases.

There was a high frequency of diagnoses for diseases of the respiratory system in classes II and III. Adult respiratory distress syndrome was the diagnosis in 8 (61.5%) of the 13 diagnoses in class II and 36 (52.9%) of the 58 diagnoses in class III; there was concordant diagnosis in 20 (27%) of 74 patients with adult respiratory distress syndrome.

In the 109 autopsy diagnoses of acute myocardial infarction, each age increase of 10 years reduced by 15% the risk of diagnostic discrepancy (OR 0.85; 95% CI 0.72 to 0.99; $p = 0.044$) and each additional day of hospitalisation increased by 3% the

Table 1 Distribution of the classes of diagnostic comparison relative to the International Classification of Diseases, 10th revision⁷

Chapter of International Classification of Diseases, 10th revision ⁷	Classes of diagnostic comparison					Total	Concordance
	I	II	III	IV	V		
Infectious and parasitic diseases	3	4	3	9	110	129	85.3%
Neoplasms	–	3	1	21	11	36	30.6%
Haematologic diseases	1	2	2	27	11	43	25.6%
Diseases of the nervous system	–	3	14	3	10	30	33.4%
Diseases of the circulatory system	28	52	86	372	1107	1645	67.3%
Diseases of the respiratory system	2	13	68	75	124	282	44.0%
Diseases of the digestive system	3	3	3	29	19	57	33.4%
Diseases of the genitourinary system	–	1	4	69	19	93	20.4%
Congenital malformations	–	1	3	32	239	275	86.9%
Haemorrhage from respiratory passages; cardiogenic, hypovolaemic and unspecified shock	–	13	7	1	215	236	91.1%
Others	4	1	7	4	42	58	72.4%
Total (%)	41 (1.4)	96 (3.3)	198 (6.9)	642 (22.3)	1907 (66.1)	2884 (100)	66.1%

risk of discrepancy (OR 1.027; 95% CI 1.002 to 1.056; $p = 0.039$). In 13 patients two diagnoses of acute myocardial infarction were made. There were differences in the time which infarctions occurred in these patients; in 10 (77%) of these 13 cases a correct diagnosis of the infarction was made at admission and in 9 (69%) the diagnosis of a second infarction was not clinically identified. There were 33 autopsy diagnoses of acute subendocardial myocardial infarction and in 22 (67%) a discrepant diagnosis was observed (12 were categorised in class III and 10 in class IV) (table 3).

Infective endocarditis was observed in 26 patients. There was concordance in the diagnosis in 19 (73%) cases and in 7 (27%) of them the clinical diagnosis was not made: one patient had a prosthetic heart valve and 6 developed native valve endocarditis. In the 19 concordant diagnoses, 10 of them were prosthetic valve endocarditis and 9 native valve endocarditis.

Pulmonary embolism was categorised in classes I and II in 30 out of 88 (34.1%) patients with this diagnosis. Pulmonary embolism was the most frequent diagnosis in classes I and II (table 3).

Diagnoses of congestive heart failure, cardiomyopathy, valvular heart disease and renovascular hypertension relative to the classes of diagnostic comparison are shown in table 5.

DISCUSSION

We observed that 30% of the patients were categorised in classes I and II (9.1% of the patients in class I and 20.9% in class II). Our findings are in the range of 14–39% of patients in both classes I and II,^{6 12 13} and in the range of 7–16% in class I and 10–34% in class II reported in previous studies.^{1 4 14–16} In a

systematic review of 53 studies, the percentage of patients in class I reached 20.7% (median 9%) and was in the range of 4.1–49.8% (median 23.5%) for both class I and class II patients.¹⁷ Thus, the numbers we observed are consistent with other recently published studies.

It was demonstrated that the number of diagnoses in classes I and II decreased at a rate of 12.4% (95% CI 7% to 17.6%) for every 10% increase in autopsy rate.¹⁷ However, diagnoses in classes I and II were less frequent in patients who died in the intensive care units in spite of the lowest autopsy rate in these units. Overall low autopsy rates in the years we had studied (23.6% in 2001 and 22.4% in 2002) in comparison to previous reports¹⁷ may have contributed to our findings.

We found a higher number of patients in classes III (26.4%) and IV (31.8%) than previously reported (14–25% for class III and 10–21% for class IV).^{4 6 16} Our finding may be related to the criteria we followed.⁴ We did not restrict the number of diagnoses for each patient as did others.⁴ In addition, diagnoses such as atherosclerosis of the aorta may be more easily found on autopsy than a clinical diagnosis. We classified this discrepant diagnosis as class IV.

The lower rate of diagnoses in classes I and II in younger patients may be ascribed to the fact that most patients under 11 years had the diagnosis of congenital heart disease; in these patients there was a high concordance rate. This is an outstanding finding relative to past experience when non-invasive imaging methods were not routine practice in cardiology.

The increase in the percentage of class I and II diagnoses in relation to age may be due to the more frequent association of co-morbidities and sometimes atypical clinical presentation of diseases in older people.¹⁸ Our finding is in accordance with experiences that revealed a lower rate of diagnoses in classes I and II in patients younger than 30 years.⁴ In one study, patients aged between 40 and 65 years had a lower percentage of diagnoses in classes I and II than did patients younger than 40 and older than 65 years.¹ Age influence was not identified in other studies.^{12–15} These differences may also be due to the different demographic and clinical characteristics of the study samples.

The difference in the distribution of classes of diagnostic comparison was not significantly different relative to sex, as observed previously,^{1 13 14 19} a different finding from a study that reported classes I and II to be more frequent in women.⁴

The distribution of classes of diagnostic comparison relative to the duration of hospitalisation did not reveal a statistically significant difference in accordance with previous studies.^{1 4 12–15}

Table 2 Distribution of patients in classes of diagnostic comparison relative to the hospital ward where patients died

	Emergency	Intensive care units	Other wards	Total patients
Class	n (%)	n (%)	n (%)	n (%)
I	18 (11.8)	11 (6.8)	8 (8.7)	37 (9.1)
II	44 (28.7)	22 (13.7)	19 (20.7)	85 (20.9)
III	32 (20.9)	56 (34.8)	19 (20.7)	107 (26.4)
IV	46 (30.1)	50 (31.0)	33 (35.8)	129 (31.8)
V	13 (8.5)	22 (13.7)	13 (14.1)	48 (11.8)
Total	153 (100)	161 (100)	92 (100)	406 (100)

Categorisation is according to the highest class of discrepancy for each patient (class I > class II > class III > class IV > class V).

Table 3 Diseases of the circulatory system* relative to the classes of diagnostic comparison

Diagnoses	Class					Total	Concordance
	I	II	III	IV	V		
Acute myocardial infarction†	2 (2)	5 (5)	24 (12)	13 (3)	65 (54)	109 (76)	59.6% (71.1%)
Acute pulmonary oedema	2	13	8	1	20	44	45.5%
Aorta dissection	2	1	0	1	12	16	75%
Infective endocarditis	1	2	2	2	19	26	73.1%
Pulmonary embolism	13	17	12	15	31	88	35.2%

*Frequency higher than 15 cases and at least one diagnosis in class I.
†Numbers between parentheses show the frequency of cases of myocardial infarction excluding subendocardial infarctions.

This finding may be understood by the fact that hospitalisation is devoted mainly to therapy, and the duration of therapy may be a function of the severity of the disease, treatment and response to therapy, and not of the diagnostic investigation.

The percentage of diagnoses in classes I and II that we observed for patients admitted to the intensive care units (20.5%) was lower than in the whole series and lower than 31.7% (53/167)¹³ and 39% (15/38)¹² reported. Several factors may contribute to our findings. Patients admitted to intensive care units may be those with clinical diagnoses that would benefit from intensive therapy including a more detailed diagnostic investigation. Patients who are severely ill or in a terminal condition may not be submitted to an extensive diagnostic investigation in a regular hospital ward due to clinical or family reasons. Such a difference in approach may be associated with the observed lower rate in diagnostic discrepancies.

The patients who died in the emergency department demonstrate the highest percentage of diagnosis in class II (28.7% in the emergency department vs 20.9% in the whole series). This finding may be understandable since patients were brought to the hospital terminally ill or even in cardiac arrest. In a previous study of a diagnostic comparison of 59 patients who died in an emergency department revealed that 4 (7%) patients were categorised in class I and 20 (34%) were categorised in class II; many of these patients were admitted to the hospital in cardiac arrest.¹⁶

In a recent study²⁰ of 255 patients who did not fulfil the clinical criteria for the diagnosis of adult respiratory distress syndrome, there were 28 (11%) patients with this diagnosis at

the autopsy. Among the 127 patients that fulfilled the diagnostic criteria of adult respiratory distress syndrome, in 43 (34%) of them the diagnosis was not confirmed at the autopsy. Both experiences demonstrate a high discordance rate and add to the challenging nature of the diagnosis of adult respiratory distress syndrome.

The diagnostic comparison of myocardial infarction was categorised in classes I and II in 7 out of 109 (6.4%) patients. In other studies there was a higher rate of diagnosis of myocardial infarction in classes I and II, in the range of 28–32%.^{1 4 14}

Among the patients who died in the intensive care units we detected 1 in 36 (2.8%) with a diagnosis of myocardial infarction in class I or II. Recent experience of patients who died in an intensive care unit¹³ also demonstrated a lower discrepancy rate in diagnoses of myocardial infarction. These findings may be related to the fact that patients referred to specialist hospitals presented with more typical symptoms and signs, hence the referral.

Increasing age was associated with a higher concordance rate for myocardial infarction. This finding may be related to the surveillance attitude of physicians caring for older people. On the other hand, the duration of hospitalisation was associated with a higher discrepancy rate, probably as a result of subendocardial infarction in critically ill patients on therapy with vasopressor drugs (eg, severely ill patients in circulatory shock). In these cases, these findings may be more properly considered a late complication of a downhill course of a critically ill patient than an additional infarction in a recovering patient, and diagnosis in this setting is usually challenging.²¹

The diagnosis of infective endocarditis was categorised in classes I and II in 3 out of 26 (11.5%) cases. The percentage was higher in another study at 25.6%.⁴ The concordance rate was higher in patients with prosthetic heart valves, recognised as an important predisposing factor that may be a diagnostic clue to infective endocarditis.²²

Pulmonary embolism was demonstrated in more than a third of patients (34.1%) in classes I and II. In fact, pulmonary embolism has been recognised as a diagnosis with a high rate of discrepancy between clinical and autopsy diagnosis: 63% in classes I and II;¹ 46.8% in classes I and II;⁴ 80.3% when the

Figure 1 Classes of diagnostic comparison I to V relative to age (classes I to IV: diagnostic discrepancy in decreasing order of importance relative to therapy and prognosis. Class V: diagnostic concordance).

Table 4 Age, sex and hospital ward where the patients died relative to the frequency of patients in classes I and II of diagnostic comparison (stepwise logistic regression)

Variable	p Value	OR	95% CI
Age (in decades)	<0.001	1.1623	1.0680 to 1.2707
Sex: male vs female	0.079	1.5018	0.9577 to 2.3787
Hospital ward: intensive care units vs emergency unit and others	0.009	0.5279	0.3240 to 0.8473

Table 5 Diagnoses of congestive heart failure, cardiomyopathy, valvular heart disease and renovascular hypertension relative to the classes of diagnostic comparison

Diagnoses	Class					Total	Concordance
	I	II	III	IV	V		
Congestive heart failure	–	–	6	13	193	212	91%
Cardiomyopathy*	–	1	1	–	31	33	93.9%
Valvular heart disease	–	–	3	14	167	184	90.8%
Renovascular hypertension	–	–	2	–	4	6	66.7%

*Dilated, hypertrophic, eosinophilic, restrictive, alcoholic and amyloidosis.

diagnosis was considered not directly responsible for the death.²³ Nevertheless, our findings revealed the concordance rate between clinical and autopsy diagnoses for pulmonary embolism (35.2%) higher than the range of sensitivity (16–29%) for the clinical diagnosis in seven studies of cases series in over 100 patients recently reviewed.²⁴

There may be several reasons for the higher discrepancy rate for pulmonary embolism. The clinical picture of pulmonary embolism may be mild and almost asymptomatic and not easily recognised. Otherwise, embolism may be severe to the point of fatal haemodynamic compromise and be ascribed to primary heart disease; pulmonary embolism in such cases becomes a secondhand diagnostic hypothesis. Smaller pulmonary emboli may lead to severe haemodynamic compromise in severely ill patients admitted to the hospital for the treatment of cardiac decompensation; diagnosis of smaller emboli may be more difficult. In patients with heart disease, an embolus may arise in the heart and Doppler studies of lower limb veins may have less diagnostic yield to identify thrombosis. In addition, even patients who die as a consequence of pulmonary emboli arising in the heart may not reveal atrial or ventricular thrombi at autopsy. Further, cardiac thrombi are usually smaller than lower limb emboli. Lung co-morbidities such as chronic obstructive pulmonary disease may decrease the yield of diagnostic tests. A pulmonary embolism may be the cause of a cardiac decompensation leading to admission or may be a complication of hospital admission and bedrest, making the diagnosis more difficult.

Our study has limitations. The percentage of autopsies relative to the total number of deaths (23.6% in 2001 and 22.4% in 2002) may not be representative of the whole population of patients who died in the hospital. Differences between autopsied and non-autopsied cases may be related to the fact that paediatric cardiologists request autopsy more frequently for a better understanding of congenital heart defects. Otherwise, older patients are less frequently submitted to autopsy; physicians do request less autopsies and the rate of family approval may be lower. Further, we may be dealing with a difference that, in spite of being statistically significant, may not invalidate the objectives and findings of this study, because all categories of diseases were represented in the study sample.

The decision for the organs we had evaluated followed epidemiological and nosological relevance as well as operational factors in the hospital. We included patients without a written diagnostic hypothesis, mainly in patients admitted in cardiac arrest or who died in the first hours of admission. In addition, the intrinsically retrospective nature of this study includes the difficulty in the review and interpretation of data registered without a protocol, as previously emphasised.¹⁶

CONCLUSION

Factors related to the patients such as age and diagnosis, and to the care such as hospital ward influenced the distribution of diagnostic discrepancy between clinical and autopsy diagnoses.

The difference was not significant relative to sex, duration of hospitalisation or surgery. The discrepancy rate was lower than in previous studies for cardiovascular diseases, such as acute myocardial infarction, infective endocarditis and pulmonary embolism, a finding that may be related to the current study being carried out in a specialist hospital. Our findings reinforce the importance of the autopsy in providing data for the evaluation of the quality of care, teaching and research into cardiovascular diseases. The evolving character of diseases and progress in therapy may be associated with changes in the pathological findings. Knowledge of these findings may contribute to the evaluation of new medical interventions. Further, important clinical hypotheses may arise from autopsy findings.

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IMAGES IN CARDIOLOGY

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Bridge to recovery in a patient with Churg–Strauss myocarditis by long-term percutaneous support with microaxial blood pump

Allergic granulomatous angiitis depicts the pathological basis of the so-called Churg–Strauss syndrome, an autoimmune process characterised by asthma or allergic rhinitis, symptomatic vasculitis and prominent eosinophilia. The myocardium is affected in two-thirds of cases, associated with a mortality rate >50% when the lung or gastro-intestinum is afflicted.

We report the case of a 25-year-old male patient who was referred to our institution owing to dyspnoea and angina pectoris. Chest radiography showed bilateral alveolar infiltrates. The admission nuclear magnetic resonance scan showed a diffuse left ventricular ejection fraction (LVEF) of 30%, and diffuse late enhancement of myocardium 15 minutes after

contrast dye (arrow, panel A). Besides increased troponin levels there was an eosinophilia of 50% in the blood cell count. Within the next 2 days, the LVEF worsened to <10%. Endomyocardial and colon biopsies showed eosinophilic granulomas, confirming the diagnosis of Churg–Strauss syndrome. Treatment comprising methyl prednisolone and cyclophosphamide was started. Owing to increasing blood lactate levels despite inotropic medication, we decided to implant an intra-aortic balloon pump (IABP). Organ hypoperfusion worsened further, as indicated by increasing lactate levels. The patient was intubated and mechanically ventilated, with development of pulmonary oedema (Killip class IV).

A microaxial blood pump (Impella Recover LP 2.5) was implanted through the left femoral artery after removal of the IABP (panel B). We set the pump speed to maximum, generating a blood flow of 2.5 l/min from the left ventricle to the ascending aorta. Blood lactate concentration decreased from 8.0 mmol/l to 1.4 mmol/l within 12 hours of circulatory support.

We were able to wean the patient from the microaxial blood pump within 6 days after implantation. No signs of leg ischaemia occurred during that period. The patient was discharged from the intensive care unit after 13 days, and from hospital after another 3 weeks, with an LVEF of 45%. This case demonstrates the ability of microaxial blood pumps to support patients with severe cardiogenic shock sufficiently if other measures, such as inotropic drugs and IABP, fail.

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