

## Cerebral abscess as a neurosurgical emergency in Eisenmenger syndrome: illustrative case

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**BACKGROUND** Eisenmenger syndrome caused by severe pulmonary arterial hypertension in congenital heart disease can cause multisystemic involvement and is a risk factor for development of cerebral abscess. Cerebral abscess, if not detected and treated in a timely manner, can present as acute neurosurgical emergency.

**OBSERVATIONS** The authors reported a case of cerebral abscess in a patient with Eisenmenger syndrome. The patient presented with acute neurological deterioration with mass effect and cerebral infarcts and received emergency neurosurgical intervention. A further literature search was done to identify prior reported cases of cerebral abscess with Eisenmenger syndrome.

**LESSONS** Patients with Eisenmenger syndrome have compromised cardiorespiratory status, and decision for neurosurgical intervention should be given careful consideration. Multidisciplinary team management along with preoperative optimization of the patient should be used.

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**KEYWORDS** cerebral abscess; Eisenmenger syndrome; congenital heart disease

Eisenmenger syndrome is a life-threatening, severe form of pulmonary arterial hypertension associated with congenital heart disease (CHD).<sup>1</sup> Eisenmenger syndrome can have multisystemic involvement, causing cerebrovascular accidents (CVAs), cerebral abscess, hemoptysis, arrhythmia, and syncope.<sup>2</sup> It has been defined as a syndrome in which “a congenital heart defect that initially causes a major left to right shunt induces severe pulmonary vascular disease and pulmonary arterial hypertension and finally results in reversal of the direction of shunting and development of cyanosis.”<sup>3</sup> Patients with Eisenmenger syndrome are prone to develop CVAs and cerebral abscess as the development of right-to-left shunt bypasses the filtering effect of pulmonary microcirculation. Cerebral abscesses, if not detected and treated early, may present as acute neurosurgical emergency and may require emergency surgery with a substantial risk of intra- or perioperative adverse events. Herein, we present the case of a young woman with cerebral abscess and Eisenmenger syndrome who had a severe, acute neurosurgical emergency.

### Illustrative Case

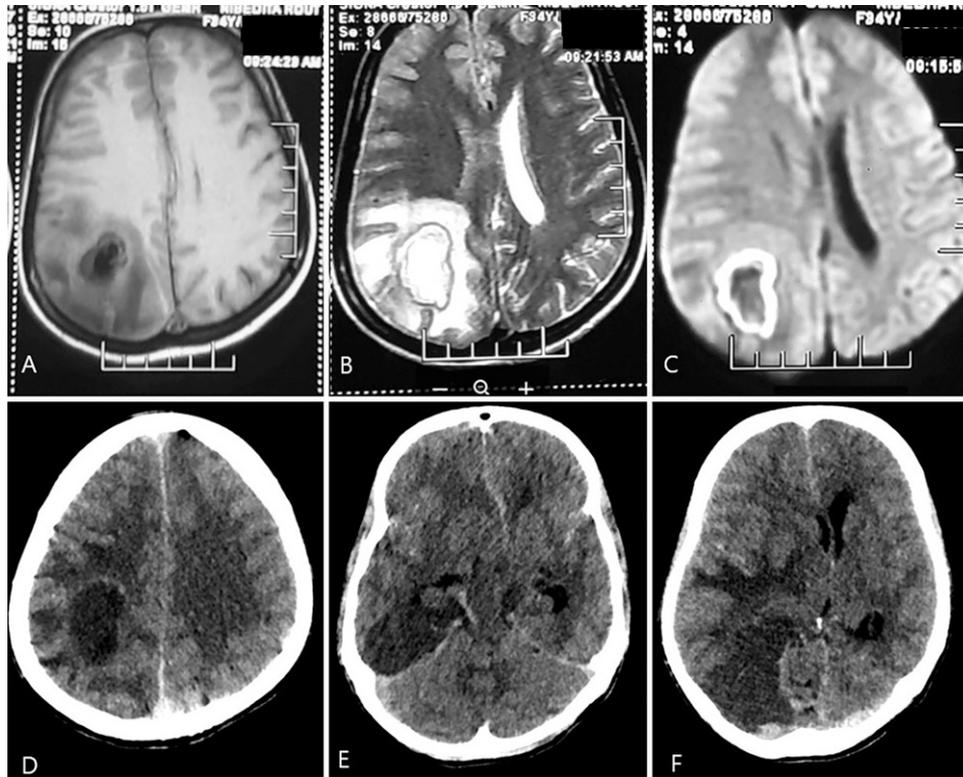
A 34-year-old married woman presented to the casualty department of our institution with history of altered sensorium followed by obtundation for 1 day. She had history of intermittent episodes of vomiting with headache for the last month that were investigated, and she had been advised on treatment at an outside hospital but was lost to follow-up. She had one child 12 years of age who had been delivered vaginally and uneventfully. She had no history of fever or limb weakness or any history suggestive of cranial nerve involvement. She did not have any history of dyspnea on exertion or cyanotic spells. On examination, her Glasgow Coma Scale (GCS) score was E1V1M2, with decerebrate posturing on stimulation. She had anisocoria with dilated, fixed right pupil. Her pulse, blood pressure, and SpO<sub>2</sub> on admission were 68/min, 102/80 mm Hg, and 85% on room air, respectively. She had no cyanosis or clubbing. She was intubated and put on mechanical ventilation, and resuscitation was started. Magnetic resonance imaging (MRI) of the brain performed at an outside institution 2 weeks earlier had shown a

**ABBREVIATIONS** ASD = atrial septal defect; CHD = congenital heart disease; CT = computed tomography; CVA = cerebrovascular accident; GCS = Glasgow Coma Scale; MRI = magnetic resonance imaging; SOL = space-occupying lesion.

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**FIG. 1. A:** T1-weighted MRI showing well-defined hypodense SOL in right posterior parietal region. **B:** T2-weighted MRI showing the SOL to be hyperintense with significant perilesional edema. **C:** Diffusion-weighted image showing the SOL with a well-defined margin with restricted diffusion, with the central part having no restriction of diffusion. **D:** Noncontrast CT scan showing a well-defined hypodense SOL in right posterior parietal region. **E:** Noncontrast CT scan showing a well-demarcated hypodense area in right temporal region suggestive of acute infarct in the inferior division of the right middle cerebral artery. **F:** Noncontrast CT scan showing well-demarcated hypodense area in posterior parietooccipital region suggestive of acute infarct in right posterior cerebral artery territory.

well-defined T1 hypointense, T2 hyperintense space-occupying lesion (SOL) in the right posterior parietal region with significant perilesional edema and mass effect (Fig. 1A–C). Noncontrast head computed tomography (CT) performed at presentation showed a well-defined hypodense SOL in the right posterior parietal region with mass effect (Fig. 1D). An acute infarct was present in the right posterior cerebral artery territory and in the inferior division of right middle cerebral artery (Fig. 1E and F). Her chest radiograph showed cardiomegaly and prominent pulmonary trunk (Fig. 2B).

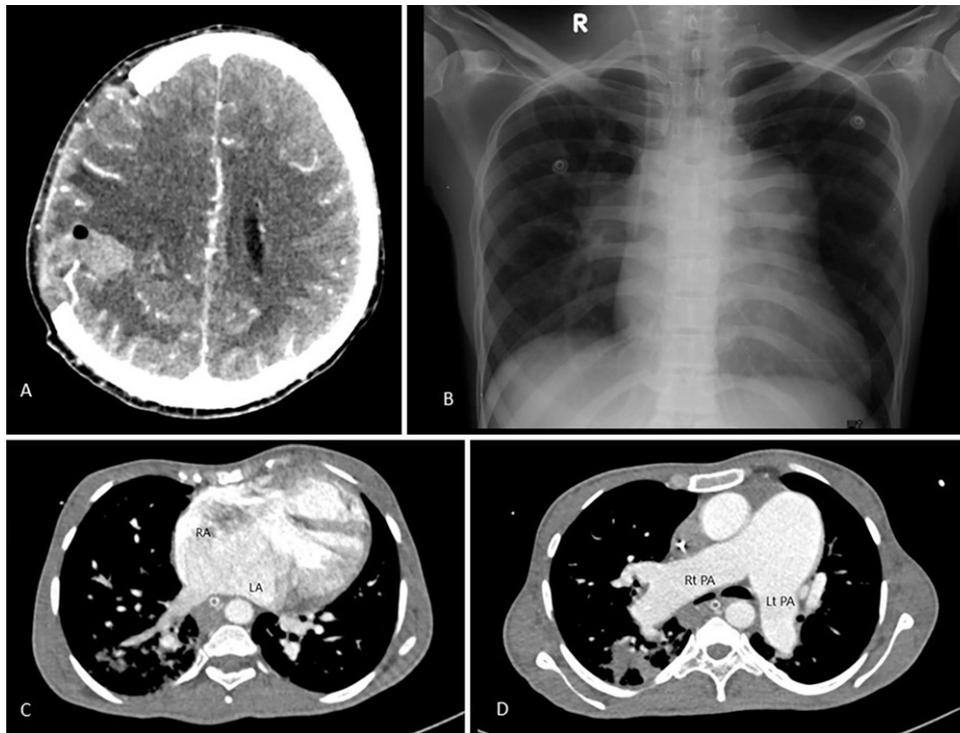
Because of signs of herniation and the presence of an infarct, she received emergency surgery. A right frontotemporoparietal decompressive craniectomy was performed along with evacuation of a right posterior parietal abscess and excision of the abscess cavity. Approximately 100 mL of pus within a thick-walled abscess cavity was present. Intraoperatively, her saturation was 92%, with an  $\text{FiO}_2$  of 80%. She required noradrenaline and adrenaline support intraoperatively. Subsequent echocardiography in the immediate postoperative period showed the presence of a large atrial septal defect (ASD), with Eisenmenger syndrome, dilated pulmonary arteries, and prolapsed anterior leaflet of the mitral valve without the presence of any vegetations. CT pulmonary angiography showed the presence of dilated main pulmonary artery (4.77 cm), right pulmonary artery (3.0 cm), and left pulmonary artery (3.1 cm) with a large ASD (Fig. 2C

and D). There was no evidence of pulmonary embolism or thrombosis. The evacuated pus showed growth of *Staphylococcus epidermidis* sensitive to ciprofloxacin, levofloxacin, vancomycin, linezolid, and tigecycline. Antibiotics were tailored accordingly. She required long-term ventilatory and inotropic support. She was tracheostomized because of long-term ventilatory requirement. She was managed by a multidisciplinary team of neurosurgeons, neuroanesthesiologists, and cardiologists. She was started on ambrisentan, tadalafil, and digoxin. Contrast brain CT obtained in the postoperative period showed complete excision of the abscess (Fig. 2A). However, she did not respond to therapy and required escalating doses of multiple inotropes. She also had poor oxygenation secondary to Eisenmenger physiology. She developed acute renal failure and succumbed to refractory hypotension on postoperative day 15.

## Discussion

### Observations

Cerebral abscess in Eisenmenger's syndrome is a rare presentation. Hence, we performed a literature search to identify previous reported cases in the literature. For literature review, we planned to include all studies meeting inclusion criteria, including case series and case reports. Literature search was carried out using the PubMed interface for all published reports in the English language using the



**FIG. 2. A:** Postoperative contrast CT scan showing complete excision of abscess and craniectomy status. **B:** Chest radiograph showing markedly prominent pulmonary trunk. **C:** CT pulmonary angiography showing the presence of large ASD. LA = left atrium; RA = right atrium. **D:** CT pulmonary angiography showing marked dilatation of right and left pulmonary arteries, marked as Rt PA and Lt PA, respectively.

following terms in combination: “cerebral abscess,” “brain abscess,” “intracranial abscess,” “Eisenmenger syndrome” (Supplementary Appendix). Inclusion criteria were human studies in which patients with Eisenmenger syndrome and cerebral abscess were reported. We extracted the following data: age, sex, type of CHD, symptoms at presentation, GCS at presentation, type of presentation (emergency/nonemergency), location of abscess, organism isolated (if any), management, and outcome (mortality). If any of the data were not available in a particular study, it was recorded as “not reported” and not included in the analysis.

A total of 10 studies fulfilling the inclusion criteria were included for analysis. Out of 10, 8 were case reports and 2 were retrospective cohort studies, making a total of 25 patients reported in the literature who had Eisenmenger syndrome and cerebral abscess (Table 1).<sup>4–13</sup> The retrospective cohort study by Daliento et al. included a cohort of patients with Eisenmenger syndrome, 7 of whom had cerebral abscess. They commented that although cerebral abscess in patients with Eisenmenger syndrome had significant morbidity, they did not affect mortality. Further clinical details were not available. Vimala et al. studied a retrospective cohort of patients with cerebral abscess, 10 of whom had Eisenmenger syndrome. Further clinical details were not available. Of the 8 case reports, 6 were males and 2 were females. Age varied from 8 to 47 years. Ventricular septal defect was the most common underlying cardiac anomaly reported, with one case of tetralogy of Fallot and another case of single atrium with patent ductus arteriosus. Headache and focal neurological signs were the most common presenting features. Most of the abscesses reported were on the left side. The organism

isolated was reported in 3 cases: 2 were *Streptococcus anginosus* and 1 was *Propionibacterium propionicum*. Of the 8 case reports, 2 received medical management, there was 1 case of craniotomy and excision of abscess, craniotomy and abscess drainage were reported in 3 cases, and stereotactic drainage was reported in 2 cases. None of the reported 8 cases had mortality.

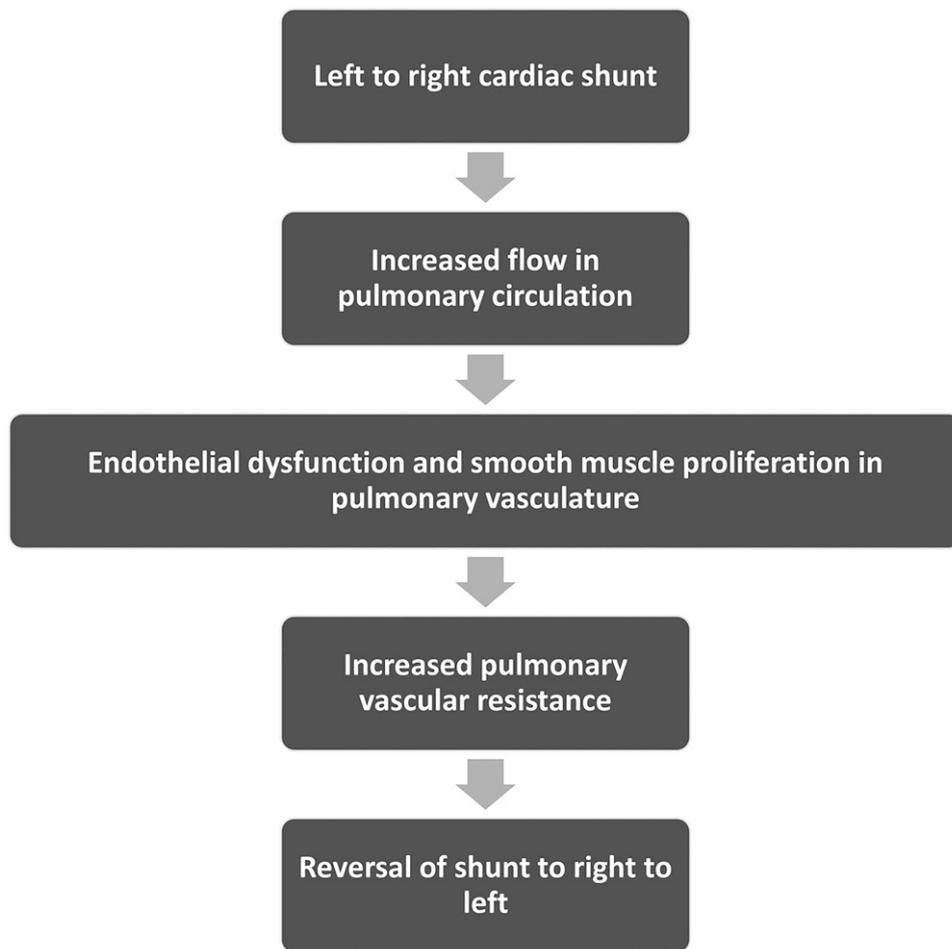
In the present era, early detection and timely repair of CHD prevents progression to Eisenmenger syndrome. In European CHD cohorts, the incidence of Eisenmenger syndrome varies from 1% to 5.6%.<sup>14,15</sup> However, once a patient develops Eisenmenger syndrome, he or she is at significant risk of developing CVAs and cerebral abscesses. In their cohort of 402 patients with brain abscesses, Vimala et al. found that 34 patients had congenital cyanotic heart disease.<sup>5</sup> Of these 34 patients, 10 had Eisenmenger syndrome. Daliento et al. reported a retrospective cohort of 188 patients with Eisenmenger syndrome, of whom 7 (3.7%) developed a cerebral abscess.<sup>12</sup>

In Eisenmenger syndrome, initially, the presence of a left-to-right cardiac shunt causes increased flow in the pulmonary circulation. This eventually leads to endothelial dysfunction and smooth muscle proliferation in the pulmonary vasculature, causing an increase in pulmonary vascular resistance. The increase in pulmonary vascular resistance causes reversal of the shunt to right to left, causing Eisenmenger syndrome (Fig. 3). This mixing of venous and arterial blood in the systemic circulation results in hypoxemia and cyanosis, causing erythrocytosis and increased blood viscosity. The right-to-left shunt also allows venous blood to bypass the filtering effect of pulmonary capillaries and go directly to the cerebral circulation. These

**TABLE 1. Included studies and patient details of cerebral abscess patients with Eisenmenger syndrome**

Authors & Year	Type of Study	No. of Subjects	Age (yrs)	Sex	Type of CHD	Sxs at Presentation	GCS Score at Presentation	Type of Presentation	Location of Abscess	Organism Isolated	Management	Outcome
Chen & Jiang, 2021 <sup>4</sup>	Case report	1	47	M	Vent septal defect	HA, memory decline	NR	Nonemergency	Lt temporal	No growth	Lt temporal craniotomy & excision of abscess	Complete neurological recovery
Vimala et al., 2020 <sup>5</sup>	Retro	10	NR	NR	NR	NR	NR	Other	NR	NR	NR	NR
Wang et al., 2019 <sup>6</sup>	Case report	1	35	F	Single atrium, patent ductus arteriosus	Fever, HA, cough	NR	Nonemergency	Lt temporal	NA	Medical management	Discharged
Casabella et al., 2019 <sup>7</sup>	Case report	1	38	M	Vent septal defect	Rt arm & face paresthesia, focal Sz of rt upper limb	NR	Emergency	Lt frontotemporal	<i>Streptococcus anginosus</i>	Stereotactic drainage	Discharged
Beachey & Wames, 2019 <sup>8</sup>	Poster	1	39	M	Vent septal defect	Rt arm weakness, paresthesias, & rt facial twitching	NR	NR	Lt parietal	<i>Streptococcus anginosus</i>	Stereotactic aspiration	Discharged
Hall et al., 2016 <sup>9</sup>	Case report	1	40	F	Tetralogy of Fallot	HA, neck pain, intermittent vomiting, & fever	NR	Emergency	Lt occipital	NA	Medical management	Discharged
Srinivasaiah et al., 2016 <sup>10</sup>	Case report	1	8	M	Vent septal defect	Fever, HA, vomiting, seizure	15	Nonemergency	Lt parietooccipital	NR	Craniotomy & evacuation of abscess	Stable
Chau et al., 2012 <sup>11</sup>	Case report	1	33	M	Vent septal defect	HA, blurred vision, expressive dysphasia, unintentional weight loss	NR	Emergency	Rt frontal	<i>Propionibacterium propionicum</i>	Craniotomy & evacuation of abscess under stereotactic guidance	Discharged
Daliento et al., 1998 <sup>12</sup>	Retro	7	24.1 ± 4.9	NR	NR	NR	NR	Other	NR	NR	NR	NR
Chaney, 1992 <sup>13</sup>	Case report	1	36	M	Vent septal defect	NR	NR	Nonemergency	Lt temporo-parietal	NR	Craniotomy & evacuation of abscess	Discharged

Ha = headache; NA = not applicable; NR = not reported; retro = retrospective; Sxs = symptoms; Sz = seizure; vent = ventricular.



**FIG. 3.** Flowchart showing the development of Eisenmenger syndrome in CHD.

factors contribute to the development of cerebral abscess in Eisenmenger syndrome.<sup>1,9</sup>

Transthoracic echocardiography is the first-line tool to evaluate a patient with suspected Eisenmenger syndrome. It helps to delineate the underlying cardiac defect, determine direction of the shunt, assess the ventricular function, and measure the pulmonary artery pressures. Transesophageal echocardiography can be performed when the transthoracic window is poor. Cardiac MRI can be used to assess the ventricular volume, assess pulmonary blood flow and assess the pulmonary to systemic flow ratio.<sup>16</sup> The gold standard to diagnose Eisenmenger syndrome is right heart catheterization because it can directly assess the hemodynamics and measure mean pulmonary artery pressure and pulmonary vascular resistance.<sup>16</sup>

Cerebral abscess is diagnosed by contrast MRI, which typically shows a well-defined ring-enhancing SOL with central T1 hypointensity, T2 hyperintensity, and restriction of diffusion. MR spectroscopy shows presence of lactate peak. In the emergency setting, contrast CT can be done to show the presence of a ring-enhancing, well-defined SOL with central hypodensity.<sup>17</sup> Dental assessment and examination of ear should be performed to look for nidus of infection. Blood cultures are indicated for systemic infections or to diagnose bacterial endocarditis.

Patients with an abscess <2.5 cm in diameter without any mass effect and GCS >12 can be given a trial of medical management

with antibiotics and dexamethasone, but abscesses not meeting these criteria should be managed surgically.<sup>17</sup> The surgical options are open excision, burr hole aspiration, and stereotactic aspiration. However, general neurosurgical principles of management of abscess must be modified appropriately when treating patients with cerebral abscess and Eisenmenger syndrome. The precarious hemodynamic condition of the patient may not allow an extensive neurosurgical intervention or any intervention at all. Hence, the management decision in these patients should be made on a case-by-case basis, taking into consideration both the cardiopulmonary and neurological condition of the patient. When opting for surgery, perioperative volume status, pulse, and blood pressure must be meticulously managed. The direction of the blood flow across the cardiac defect is controlled by the balance between pulmonary vascular resistance and the systemic vascular resistance. For all practical purposes, in the clinical setting, pulmonary vascular resistance remains the same. However, systemic vascular resistance can vary widely during surgery. When systemic vascular resistance decreases, right-to-left shunting causes hypoxemia, which may cause further neurological deterioration. On the other hand, when systemic vascular resistance increases, there is left-to-right shunting, risking right heart failure. Maintenance of hemodynamic parameters is thus of utmost importance during surgery. Transesophageal echocardiographic monitoring should be done to look for any paradoxical emboli.<sup>13</sup>

In patients with Eisenmenger syndrome who present with cerebral abscess as an emergency, careful consideration should be given in selecting the optimal surgical procedure. Both stereotactic aspiration and surgical excision of the abscess allow for identification of the causative organism and subsequent tailoring of the antibiotic therapy. Aspiration of abscess has a low morbidity (5%) and shorter operating time, but it is associated with a postaspiration relapse rate of up to 32%.<sup>18</sup> This may necessitate multiple aspirations or excision for full resolution. Craniotomy and abscess excision have lower recurrence rates than excision; however, they are associated with prolonged operating time and greater hemodynamic changes. In patients with secondary effects such as significant mass effect, hydrocephalus, or infarct, excision is preferred.<sup>18</sup>

Cardiological management of Eisenmenger syndrome should be continued with endothelin receptor antagonists (bosentan), phosphodiesterase 5 inhibitors (sildenafil, tadalafil), and prostacyclin analogs in consultation with cardiologists.<sup>17</sup> Iron deficiency should be looked for and corrected. The use of anticoagulation therapy in patients with Eisenmenger syndrome is controversial because it increases the risk of hemoptysis, CVA, and hemorrhage.<sup>19</sup> Heart-lung transplantation or lung transplantation with repair of cardiac defect is also a therapeutic option for patients with Eisenmenger syndrome.

In this case, presence of acute herniation with infarct prompted us to perform a decompressive craniectomy. However, it is up for discussion whether patients with Eisenmenger syndrome can tolerate such a major neurosurgical procedure with their susceptible cardiopulmonary status.

### Lessons

Patients with Eisenmenger syndrome are a fragile group of patients who require tailored therapy and meticulous surgical and anesthetic management for optimal outcome. The significant risks of major neurosurgery should be given due consideration while deciding treatment in patients with Eisenmenger syndrome. If feasible, stereotactic drainage should be considered to minimize major stress to the cardiorespiratory system. However, in the presence of mass effect and infarcts due to herniation, open decompression and excision should be considered.

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### Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

### Author Contributions

Conception and design: Sekar. Acquisition of data: Datta. Drafting the article: Datta. Critically revising the article: both authors. Approved the final version of the manuscript on behalf of both authors: Sekar. Statistical analysis: Datta. Study supervision: Sekar.

### Supplemental Information

#### Online-Only Content

Supplemental material is available with the online version of the article. *Supplementary Appendix*. <https://thejns.org/doi/suppl/10.3171/CASE2252>.

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