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## Digital subtraction angiography in evaluation of vascular supply of head and neck paragangliomas

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### Summary

**Background:**

Paragangliomas (PGs) of the head and neck are relatively rare and represent 0.6% of all head and neck tumors and 0.03% of all tumors. There are four groups of head and neck PGs: carotid body tumors, vagal PGs, jugular PGs, and tympanic PGs. The resection of head and neck PGs carries an inherent risk of injury to cranial nerves and vascular structures which may lead to excessive bleeding. To plan the surgical strategy for PGs, detailed information about the vascular supply of the tumor is required.

**Material/Methods:**

Between January 1998 and April 2007, 41 tumors of the head and neck were identified in 37 patients (20 females, 17 males, mean age: 38.4 years). Single tumors were observed in 33 patients, two head and neck PGs were identified in 3 patients, and 1 patient presented 3 PGs, one of which was located laterally to the aortic arch. There were 21 PGs located at the carotid bifurcation, 10 in the jugular foramen, 6 in the tympanic cavity, and 4 along the course of the vagus nerve.

**Results:**

In all the cases of PGs located in the head and neck, the vascular supply came from branches of the external carotid artery. Vascular supply from the internal carotid and the vertebral arteries was not seen in any of the patients. The most common vascular supply in the cases of carotid body tumors and jugular PGs was the pharyngeal ascending artery. In the cases of vagal PGs it was the pharyngeal ascending artery and the posterior auricular artery and in the case of tympanic PGs the posterior auricular artery.

**Conclusions:**

DSA is an important tool in the diagnosis of head and neck PGs. The evaluation of its vascularization is essential in planning further treatment, both endovascular and surgical.

**Key words:**

Paraganglioma • vascularization • digital subtraction angiography • embolization

**PDF file:**

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### Background

Paragangliomas (PGs) of the head and neck are relatively rare, highly vascularized tumors which stem from neural-crest progenitor cells. According to their site of origin and location, they can be divided into the following groups: 1) carotid body tumors located at the carotid bifurcation, 2) vagal PGs located along the course of the vagus nerve in close anatomic relation to the inferior or superior ganglion, 3) jugular PGs located in the jugular foramen, and 4) tympanic PGs located in the tympanic cavity [1, 2, 3].

Other, uncommon sites have been reported, such as the nose and paranasal sinuses, the larynx, and the orbit [4, 5]. PGs represent 0.6% of all head and neck tumors and 0.03% of all tumors [6]. They are multiple in 30% of patients [7]. Familial paragangliomas have an overall prevalence of 7%-9% [3]. Less than 2% of head and neck PGs have catecholamine secretory function [8]. These PGs may be associated with familial paraganglioma, neurofibromatosis type 1, von Hippel-Lindau disease, Carney triad, and, rarely, multiple endocrine neoplasia (MEN) type 2 [9].

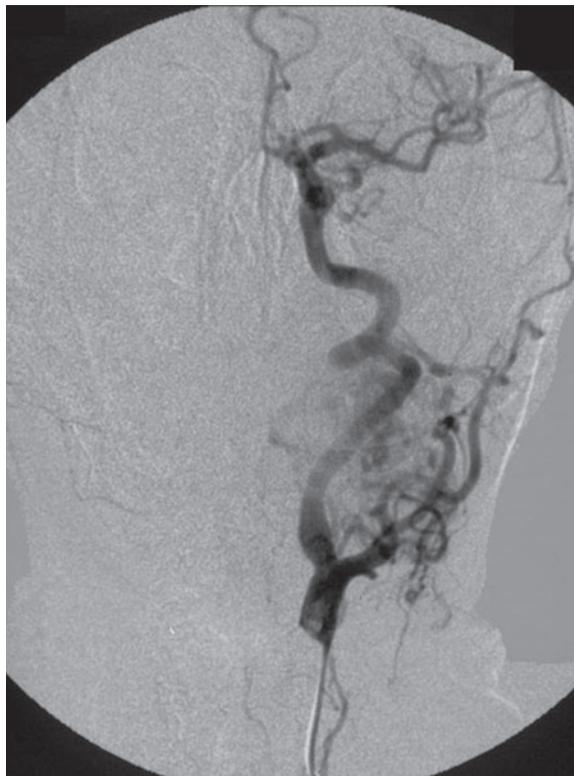


**Figure 1 A.** DSA of the aortic branches. Paraganglioma at the carotid bifurcation.

Although malignant PGs occur only in 5% of cases [10, 11], paraganglioma-associated morbidity takes place because the local growth of the tumor endangers neurovascular structures in the neck, skull base, and posterior cranial fossa. The presumptive diagnosis is made on the basis of clinical symptoms and is confirmed by imaging techniques such as computed tomography (CT), magnetic resonance imaging (MRI), or angiography. Because of their vascularity, invasive character, and critical location, these tumors remain a formidable challenge to treatment. Surgical excision has become the modern approach to these neoplasms [12, 13, 14, 15]. The resection of head and neck PGs carries an inherent risk of injury to cranial nerves and vascular structures, which may lead to excessive bleeding. In order to plan the surgical strategy for PGs, detailed information about the vascular supply of the tumor is required. Digital subtraction angiography (DSA) is the standard diagnostic method for the assessment of vascularization of PGs [7]. Preoperative embolization of tumor feeders is undertaken to reduce the hemorrhage associated with the resection of highly vascularized PGs or as a palliative treatment [16]. The aim of this study is to assess PGs' blood supply, which is essential for planning a surgical operation or preoperative embolization.

### Materials and methods

Between January 1998 and April 2007, 37 angiographies were performed on 37 patients with diagnosed paragangliomas of the head and neck at the Vascular Laboratory of the Clinical Radiology Department of University Hospital



**Figure 1 B.** The same patient. Selective angiography of the common carotid artery.



**Figure 1 C.** The same patient. Selective angiography of the ascending pharyngeal artery.

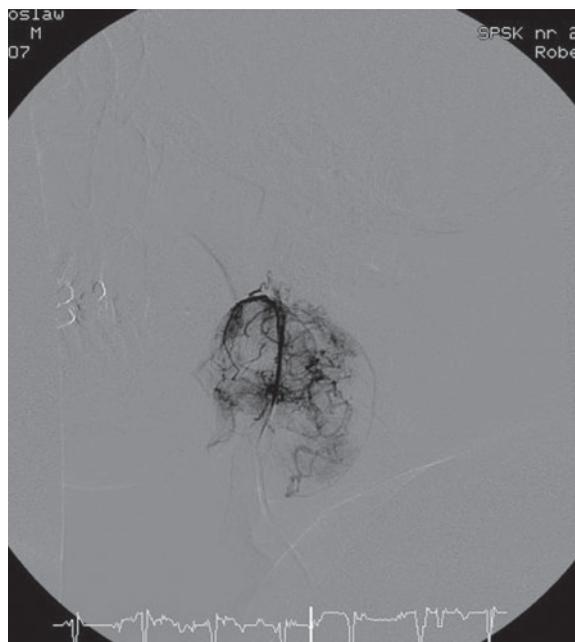
No. 1 and at the Vascular Laboratory of University Hospital No. 2 in Poznań. The subject group consisted of 20 women and 17 men aged between 24 and 65 years (mean age: 38.4). Twenty-six of them had previously had magnetic resonance imaging (MRI) performed, 22 had computed Tomography (CT), and 11 patients were diagnosed using both examination methods.



**Figure 2 A.** Selective angiography of the right common carotid artery. Paraganglioma at the carotid bifurcation.

In the 37 patients, 41 lesions located in the head and neck were identified. Single neoplasms were identified in 33 patients, tumors in two separate locations in a further three patients, and one patient had, apart from the two tumors located in her head and neck, a tumor in the mediastinum, laterally to the aortic arch. Of the 33 lesions in the head and neck in the 33 patients with single neoplasms, 20 were located at the carotid bifurcation and 6 at the jugular foramen. Lesions were identified in the middle ear in 5 cases and along the vagus nerve in 2 patients. Of the remaining 4 patients, two had lesions within the jugular foramen and along the vagus nerve. One patient had a PG at the carotid bifurcation and in the jugular foramen. In the remaining patient the PGs were located in the jugular foramen, the middle ear, and laterally to the aortic arch.

In all 37 patients, carotid and vertebral DSA were performed. DSA was performed at the Vascular Laboratory of the Clinical Radiology Department of Clinical Hospital No. 1 using a Phillips Multidiagnost apparatus with DSA as well as a Phillips Allura apparatus. However, at the Vascular Laboratory in Clinical Hospital No. 2 it was performed using a Phillips Allura apparatus. Angiography was performed under local anesthesia using Seldinger's method. After puncturing the femoral artery, a 6F introducer with a haemostatic valve (Balton, Poland) was introduced into the vessel. Then a hydrophilic guide wire was used to advance a 5F pigtail catheter (Balton, Poland) into the aortic arch. In all cases, angiography of the aortic arch was performed to determine the extent of the neoplasm's vascularization. Next, using Left Carotid, Head Hunter, and Sidewinder catheters (Balton, Poland), both common carotid arteries and vertebral arteries were catheterized. The examination was performed in two projections: p-a and lateral. In selected cases, various diagonal projections were employed



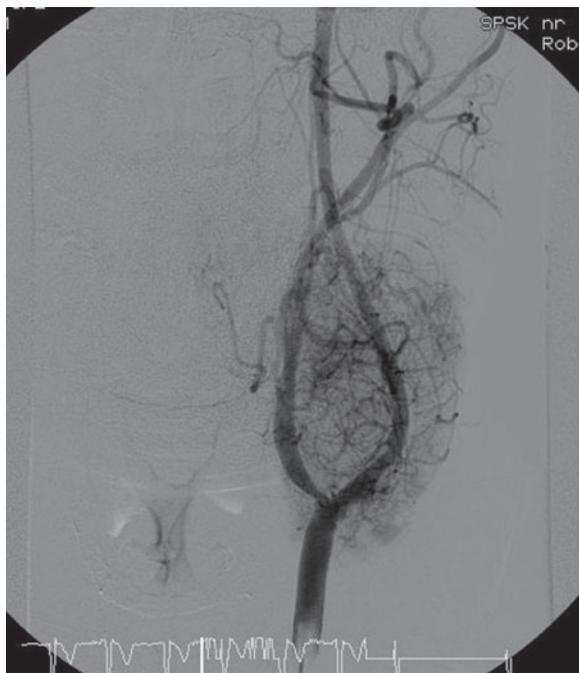
**Figure 2 B.** The same patient. Selective angiography of the ascending pharyngeal artery.

for optimal visualization of pathological vascularization. In the cases of pathological vessels arising from the common carotid artery, both the internal and external carotid arteries were always catheterized super-selectively. In the cases of elective endovascular embolization, branches of the external carotid artery were catheterized.

All angiographic examinations were performed using non-ionic contrast media, i.e. Omnipaque (Nycomed), Ultravist (Schering), and Iomeron (Altana). Angiography of the aortic arch, the common carotid arteries, and the vertebral arteries was always performed using an automatic injector which administered a specified amount of contrast medium. In the case of aortic arch angiography, 40 ml of contrast medium was administered at an outflow rate of 30 ml/s. In the cases of selective angiography of the common carotid arteries, 10 ml of contrast medium was administered at 5 ml/s. However, for angiography of the internal carotid, external carotid, and vertebral arteries, 7 ml was administered at 4 ml/s. DSA covered the early arterial phase as well as the late venous phase in order to evaluate blood flow into the veins and to determine the neoplasm's relation to the internal jugular vein. Based on DSA, the extent and range of the lesion's vascular supply, the dimensions of the pathological vessels, and the tumor's relation to the large neck vessels were determined in detail.

## Results

In all cases of paragangliomas located in the head and neck, the vascular supply came from branches of the external carotid artery. Vascular supply from the internal carotid artery and the vertebral artery was not seen in any of the patients. In all 20 cases of single tumors located at the carotid bifurcation there was evidence of splaying of the internal and external carotid arteries (Figs. 1, 2, 3). Neoplastic infiltration resulting in vessel stenosis was



**Figure 3 A.** Selective angiography of the left common carotid artery. Paraganglioma at the carotid bifurcation.

not observed in any of the cases. In all cases, the neoplasm caused lateral displacement of the jugular vein. The vascular supply in 18 cases came from the ascending pharyngeal artery, in 14 cases from the superior thyroid artery, in 9 cases from the lingual artery, in 8 cases from the facial artery, in 7 cases from the occipital artery, and in 5 cases from the posterior auricular artery.

Neither the carotid artery nor the internal jugular vein were displaced by neoplasms located at the jugular foramen. The vascular supply in 6 patients with single PGs located at the jugular foramen came from the ascending pharyngeal artery, in 4 patients from the posterior auricular artery and the maxillary artery, and in 2 cases from the occipital artery (Fig. 4).

Neoplasms of the middle ear displaced neither the carotid arteries nor the jugular veins. The vascular supply of the 5 middle-ear PGs came from the posterior auricular artery in 4 cases, from the ascending pharyngeal artery in 3 cases, and from the occipital artery (Fig. 5).

Two single PGs of the vagus nerve anteriorly displaced the internal and external carotid arteries and the jugular vein. Both were supplied by the ascending pharyngeal artery and the posterior auricular artery (Fig. 6). In one case the vascular supply came from the facial and the occipital arteries. In both patients with PGs of the jugular foramen and the vagus nerve, slight anterior displacement of the neck's blood vessels was observed. The vascular supply came from the ascending pharyngeal artery, the posterior auricular artery, and the occipital artery. In the patient with PGs located at the carotid bifurcation and at the jugular foramen, the vascular supply came from the ascending pharyngeal artery, the maxillary artery, and the posterior auricular artery. Lateral displacement of the jugular vein and splaying of the



**Figure 3 B.** The same patient. Selective angiography of the external carotid artery.

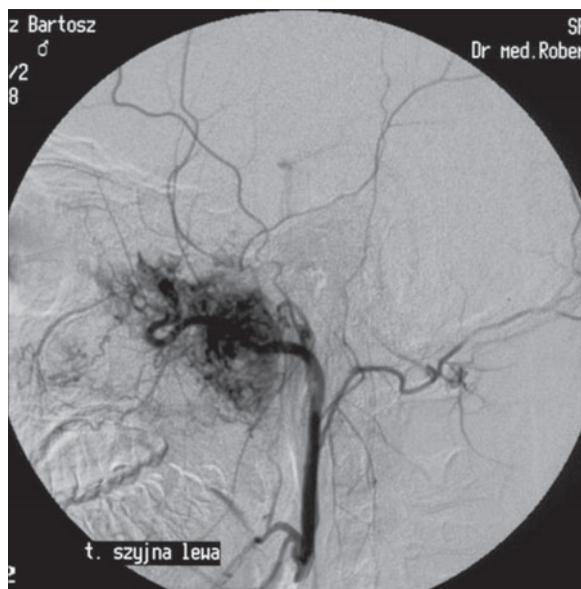


**Figure 3 C.** The same patient. Selective angiography of the ascending pharyngeal artery.

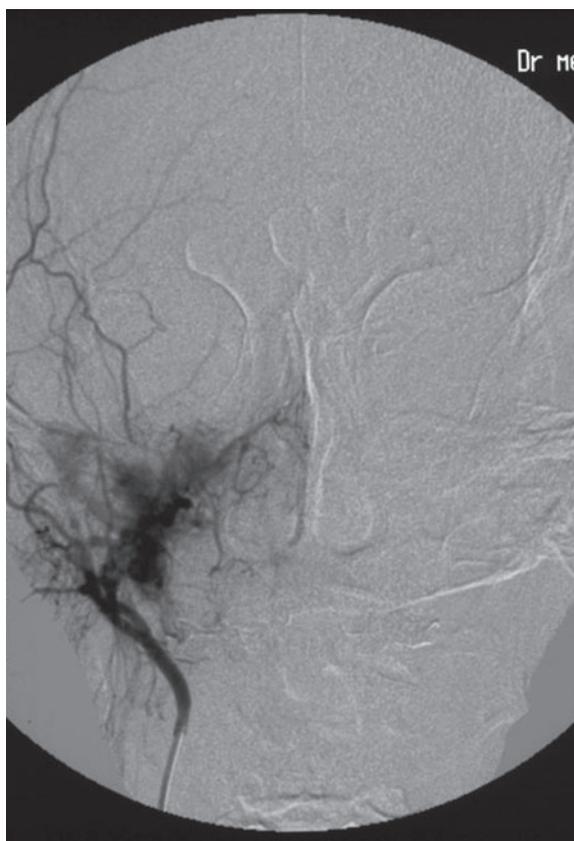
carotid bifurcation were observed. In the patient with PGs located in three places, the vascular supply for the tumors in the head and neck came from branches of the posterior auricular artery, the ascending pharyngeal artery, and the maxillary artery. In her case, no displacement of the neck's vessels was observed. The vascularization of the head and neck paragangliomas is summarized in Table 1.

## Discussion

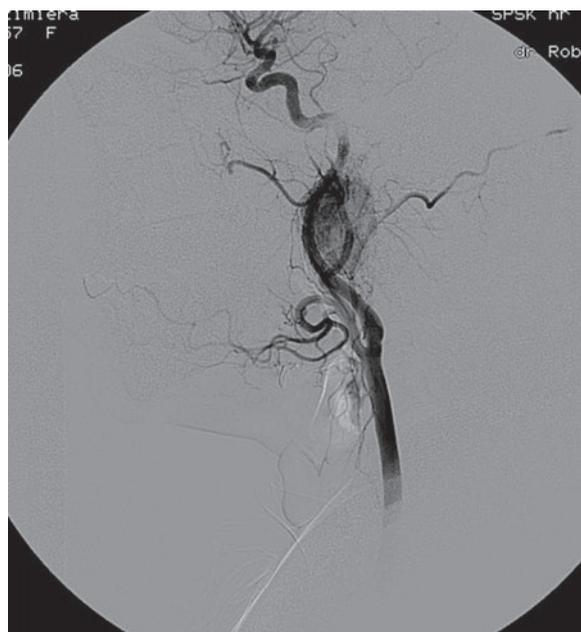
For practical reasons, angiography combined with endovascular treatment of head and neck pathology can be divided into three main groups: 1) angiography as a typically diagnostic examination which allows us to diagnose and to determine



**Figure 4.** Selective angiography of the left external carotid artery. Glomus jugulare.



**Figure 5.** Selective angiography of the right external carotid artery. Glomus tympanicum.



**Figure 6 A.** Selective angiography of the right common carotid artery. Glomus vagale.

the area supplied by a particular vessel and the main vascular source, 2) angiography with embolization as a diagnostic as well as therapeutic examination which fully eliminates the pathology that is the cause of hospitalization (recurrent epistaxis, angioma) [17], often referred to as radical embolization [18] or palliative embolization of benign and malignant neoplasms as a basic therapeutic procedure [19], and 3) angiography with embolization as a supportive procedure before elective surgery aimed at the restriction of bleeding [20].

Angiography is a technique and diagnostic method which enables the determination of anatomical conditions, preliminary verification of the diagnosis, as well as, from the



**Figure 6 B.** The same patient. Selective angiography of the ascending pharyngeal artery.

surgeon's standpoint, preparation of a suitable plan for the surgical procedure [21]. The result of the angiographic examination determines the extent of the procedure and what steps should be taken during surgery to prevent excessive bleeding. Tumors supplied by the external carotid

**Table 1.** Vascularization of paragangliomas.

Location	Number	Vascularization	Relationship to the jugular vein	Relationship to the carotid arteries
single carotid body tumor	20	ascending pharyngeal a. – 18 superior thyroid a. – 14 lingual a. – 9 facial a. – 8 occipital a. – 7 posterior auricular a. – 5	displacement (n=20)	displacement (n=20)
single jugular PG	6	ascending pharyngeal a. – 6 maxillary a. – 4 posterior auricular a. – 4 occipital a. – 2	no displacement (n=6)	no displacement (n=6)
single tympanic PG	5	posterior auricular a. – 4 ascending pharyngeal a. – 3 occipital a. – 3	no displacement (n=5)	no displacement (n=5)
single vagal PG	2	ascending pharyngeal a. – 2 posterior auricular a. – 2 facial a. – 1 occipital a. – 1	displacement (n=2)	displacement (n=2)
multiple in 4 patients	8	ascending pharyngeal a. – 4 posterior auricular a. – 4 occipital a. – 2 maxillary a. – 2	displacement (n=6) no displacement (n=2)	displacement (n=6) no displacement (n=2)

PG – paraganglioma; a. – artery

artery are more eagerly embolized because there is less risk of dangerous complications, especially those resulting from flushing the thrombus or embolus into the central nervous system. Occlusion of the internal carotid artery's branches is also performed, despite the substantial risk of complication (occurrence of micro-embolisms, ischemic strokes, or sudden blindness resulting from occlusion of the ophthalmic artery) [22]. This procedure is combined with special preparation of the patient (heparinization) as well as selection of suitable embolization agents. The above technique is widely used in the embolization of brain tumors and arteriovenous malformations [23], though it is also used in the embolization of neck tumors. In analyzed material comprising 41 paragangliomas of the head and neck, the vascular supply was solely from the external carotid artery, whereas no vascular supply from the internal carotid and vertebral arteries was observed.

Patients with paragangliomas of the head and neck make up a group of patients in whom angiography combined with embolization is applied as preparation for surgical removal of the tumor (as a supporting procedure before elective surgery). This procedure aims to reduce intraoperative hemorrhage, an important factor in terms of the patient's health (no need to refill the vascular bed, no need to infuse blood supplements) as well as improvement of the operation's safety [15]. Angiography also provides valuable information on the location of blood vessels. In 21 patients with carotid glomus (glomus caroticum), typical divergence of the internal and external carotid arteries at their bifurcation was observed as well as lateral displacement of the internal jugular vein, a valuable clue for the surgeon. Also important is the fact that no signs of infiltration of the vascular wall were observed. In 18 tumors of the carotid glomus, the

vascular supply was from the ascending pharyngeal artery, which sometimes originates directly from the vicinity of the carotid bifurcation. Displacement of blood vessels was also not observed in the group of patients with tympanic and jugular glomera. Minor anterior displacement of blood vessels was observed in two cases of vagus nerve paragangliomas as well as in two patients with paragangliomas of the foramen jugular and the vagus nerve. Elective surgery should be carried out four to ten days after embolization. This is due to recanalization, secondary clearing of the blood vessels, which in its final effect does not allow for the achievement of the primary goal, which is the reduction of bleeding. Angiography combined with embolization as a preparatory procedure is applied in cases of paragangliomas of the carotid bifurcation (glomus caroticum), paragangliomas of the bulb of the jugular vein (glomus jugulare), PGs of the vagus nerve (glomus vagale), and PGs developing in the middle ear (glomus tympanicum) [24]. In some cases of large tumors penetrating the skull's interior (glomus jugulare, vagale, tympanicum) as well as the case of general burdens to the patient which make it impossible to operate, angiography combined with embolization, along with successive irradiation, is considered palliative therapy which hampers the tumor's further development [24]. During embolization of the glomus tympanicum and some cases of the glomus jugulare and vagale, we must take into account the possibility of peripheral paresis of the facial nerve due to the embolization agent flowing into the stylomastoid ramus of the posterior auricular artery.

Angiography, however safe it may be considered, is not free of complications. Among the most serious complications are death as well as complications from the central nervous system resulting from the above-mentioned risk

of thrombus or embolus flushing. One must also keep in mind the occurrence in some patients of the so-called post-embolization syndrome. This is a group of symptoms comprising fever, nausea, vomiting, pain, and elevated white blood cell count [3]. They are attributed to tumor ischemia, are temporary, and may indicate that the treatment was successful [3]. These symptoms appear shortly after surgery and normally recede after 3-5 days. This syndrome is linked to reaction to the occlusive agent and also depends on the anatomical location and technique of administering the agent. On its own, post-embolization syndrome is not life-threatening and is treated conservatively. However, it should be differentiated from conditions resulting from other somatic disorders, such as local infection or increased intracranial pressure. Major complications include cerebral ischemia. This occurs as a result of an accidental introduction of embolic material into the vertebrobasilar system

through the external carotid artery or through the existing anastomoses between the external carotid artery and the internal carotid artery [3,25]. The anastomoses between the external carotid artery and the vertebrobasilar system are formed by the superior thyroid artery and the occipital artery [26].

All in all, angiography combined with embolization plays an important role in the treatment of the head and neck tumors. As a diagnostic tool it provides information on the extent and range of vascularization as well as the location of major vessels of the head and neck. However, augmented by embolization it becomes an element of a therapeutic procedure which results in a major improvement of the conditions under which surgery is performed. In select cases, embolization may be an alternative to surgical intervention.

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